3.1 OESOPHAGUS

HIATUS HERNIA

DEFINITION

- Protrusion of part of the stomach through the diaphragmatic oesophageal opening
  - **Sliding hernia** (the commonest type): the gastro-oesophageal junction (GOJ) slides proximally through the diaphragmatic hiatus to assume an intrathoracic position ► it is accompanied by reflux and oesophagitis
    - The squamocolumnar junction is seen at ≤38cm (the normal is 40cm) from the incisors at endoscopy
  - **Rolling hernia:** the GOJ is in a normal position below the diaphragm – the proximal stomach (usually the fundus) herniates through the hiatus ► this is more prone to incarceration and obstruction, and it may undergo torsion, resulting in strangulation, infarction or perforation
    - The squamocolumnar junction maintains its normal position
  - **Combined hernia:** features of both are present

CLINICAL PRESENTATION

- Asymptomatic or gastro-oesophageal reflux (± reflux oesophagitis) ► symptoms are more commonly seen with a sliding hernia

RADIOLOGICAL FEATURES

Barium swallow

- **Sliding hiatus hernia:** the Schatski or B ring is demonstrated above the diaphragmatic hiatus
- **Rolling hiatus hernia:** a part of the stomach (usually the gastric fundus) is prolapsed into the chest anterior or lateral to the oesophagus

PEARLS

- **Schatski or B ring:** a ring of mucosal tissue at the lower border of the phrenic ampulla marking the junction between the squamous and columnar epithelium (the 'Z line')
- *The 'A' ring or inferior oesophageal sphincter:* about 2-4cm proximal to the B ring is a thicker ring produced by active muscular contraction
- The Schatski ring is always associated with a small sliding hiatus hernia ► it can be congenital or secondary to gastro-oesophageal reflux (with associated inflammation and fibrosis)
- The Schatski ring is usually no more than 2–3 mm in thickness ► despite being mucosal it can be symptomatic (requiring dilatation)
- If the B ring is incomplete, part of it can sometimes be demonstrated as the incisural notch (which is inevitably seen on the greater curve aspect of the stomach)

GASTRO-OESOPHAGEAL REFLUX DISEASE (GORD)

DEFINITION

- GORD follows lower oesophageal sphincter dysfunction ► this initially leads to reflux (with minor irritation and inflammation) but can then proceed to ulceration, fibrosis and stricture formation ► it may also be associated with a hiatus hernia

CLINICAL PRESENTATION

- Heartburn or dysphagia ► the major long-term complications are peptic oesophagitis (± stricture formation or Barrett's oesophagus)

RADIOLOGICAL FEATURES

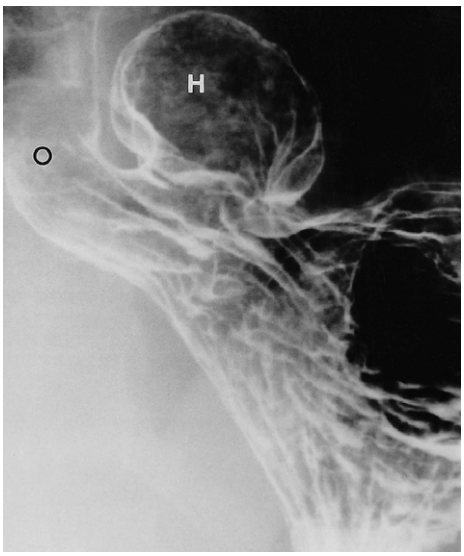
Barium swallow

- *Reflux:* this may be demonstrated but alone is of questionable significance – minor amounts can occur in the normal population ► gross reflux (up to the level of the aortic knuckle or above and not cleared by a stripping wave passing down the oesophagus) is likely to be symptomatic
  - Associated features: a wide gastro-oesophageal junction (> ⅔ of the maximally distended thoracic oesophagus) ► an inflammatory gastro-oesophageal polyp (seen as a single linear polyp straddling the GOJ)
- *Reflux oesophagitis:* this can demonstrate mucosal oedema, erosive disease or frank ulceration ► initially the collapsed oesophagus shows thickened longitudinal folds (>3mm) ► multiple fine ulcers give the mucosa a punctate or granular appearance ► larger discrete punched-out ulcers can develop ► ulceration is most pronounced immediately above the GOJ and local circular muscle spasm may produce transverse folds ► scarring produces permanent folds that radiate from the ulcer margins
- *Long-term sequelae:* stricture formation (typically a short stricture above a hiatus hernia with smooth tapered margins) ► the development of Barrett's oesophagus (in 10% of cases)

Radionuclide study Reflux of ^{99m}Tc-sulphur colloid labelled scrambled egg can demonstrate gastro-oesophageal reflux

PEARLS

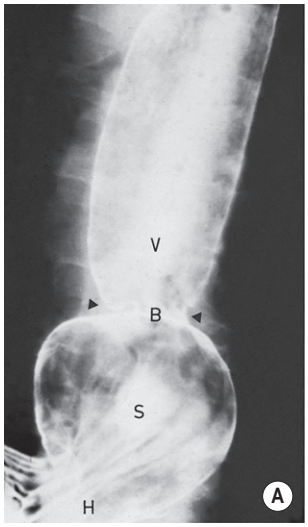
- 24-hour pH measurement is the 'gold standard' in the assessment of reflux
- There is no direct relationship between a hiatus hernia and GORD: many patients have a hiatus hernia but no GORD (but most patients with GORD will have a hiatus hernia)



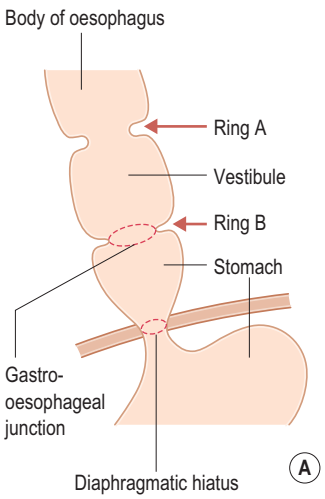
Rolling (paraoesophageal) hiatus hernia. The gastric fundus (H) lies alongside the lower oesophagus (O).†

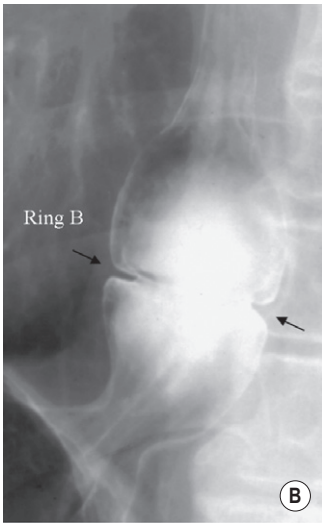
Contrast study demonstrating a combined-type hiatus hernia. Note the rolling component with a large portion of stomach above the diaphragm, but in addition the gastro-oesophageal junction has also migrated cranially.*

Inflammatory polyp (arrows) lying at end of gastric fold (asterisk).†



Sliding hiatus hernia. (A) Barium swallow shows a hiatus hernia (H), more than 3cm wide with at least 3 gastric folds seen extending across it ► S = stomach forming the hernia ► B = B ring, the gastro-oesophageal junction ► V = vestibule. The A ring is not visible. (B) CT scan showing the crura of the diaphragm (arrows) separated by 28mm (normal is <15mm). The fundus of the stomach is seen herniating through the diaphragmatic hiatus.†





The lower end of the oesophagus. (A) The B ring may normally be within 2cm above (as shown here) or below the hiatus. Thus the oesophageal vestibule may normally be above, or straddle the diaphragmatic hiatus. (B) Small sliding hiatus hernia with normal B ring (between arrows).†

OESOPHAGITIS AND BENIGN STRICTURES

DEFINITION

- Oesophageal inflammation (± subsequent smooth benign stricture formation) can be caused by the following:
  - **GORD** (see separate section)
  - **Infection:** especially in the immunocompromised patient *Candida albicans* ► herpes simplex virus (HSV) ► cytomegalovirus (CMV) ► human immunodeficiency virus (HIV) ► tuberculosis
  - **Drugs:** potassium chloride tablets ► tetracycline ► clindamycin ► doxycycline ► NSAIDs
  - **Radiation:** this is often self-limiting
  - **Crohn's disease:** this is very rare and usually accompanied by extensive GI disease elsewhere
  - **Iatrogenic:** following prolonged placement of a nasogastric tube (NGT)
  - **Caustic ingestion of strong acids or alkalis**

CLINICAL PRESENTATION

- Odynophagia ► dysphagia ► haematemesis

RADIOLOGICAL FEATURES

Barium swallow

- **Candidiasis:** initially there is dysmotility and atony of the oesophagus ► eventually classic plaque-like filling defects with ulceration and pseudomembrane formation are seen (there are also irregular and thickened mucosal folds) ► occasionally pseudo-ulcerations may appear as aphthous ulcers
- **HSV:** vesicles in the upper and mid-oesophagus appear as sessile filling defects ► when they burst they leave punched-out superficial ulcers on a background of normal mucosa ► in advanced disease there can be diffuse ulceration
- **CMV/HIV:** presents with giant oesophageal ulcers
- **Drugs:** potassium chloride causes deep ulceration leading to stricture formation ► NSAIDs can cause contact oesophagitis
- **Radiation:** > 20 Gy results in a transient oesophagitis with aperistalsis or tertiary contractions ► >45 Gy results in obliterative endarteritis after 6 months with severe oesophagitis and smooth strictures – deep ulcers can also form (which may fistulate to the trachea)
- **Crohn's disease:** this can present with aphthoid ulcers or frank ulceration
- **Nasogastric tube:** this renders the lower oesophageal sphincter incompetent, resulting in a reflux oesophagitis and a long tapered stricture within the lower oesophagus ► this may occur only 48 h post placement ► the strictures are often long and extensive
- **Caustic ingestion:** this can lead to mucosal necrosis with ulceration and mucosal sloughing ► the oesophagus may perforate within the 1st 2 weeks or result in fistulation to the pleural cavity or pericaridium ► it heals with fibrosis and stricture formation ► strictures occur at the normal sites of oesophageal compression (e.g. at the level of the aorta, left main bronchus or diaphragmatic hiatus)

PEARLS

Epidermolysis bullosa dystrophica

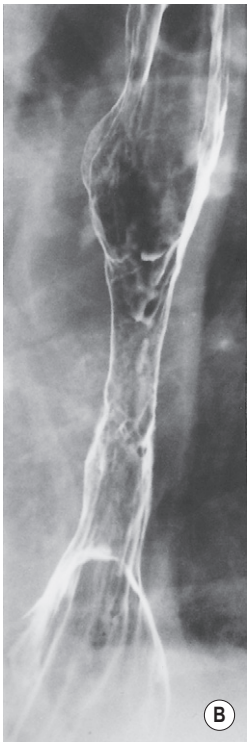
- A hereditary skin disease affecting children where minor trauma produces bullae formation ► the oesophagus may be involved (leading to stricture formation)

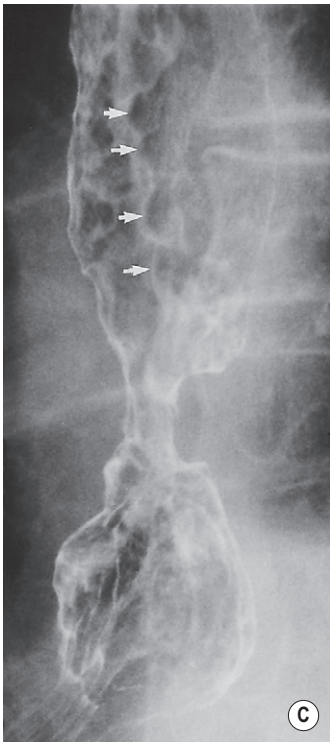
Pemphigoid

- A benign mucous membrane disease of middle age, involving the conjunctiva and mucosa of the oral cavity and skin ► the upper oesophageal mucosa may be involved with ulcers, webs and stricture formation

Intramural pseudodiverticulosis

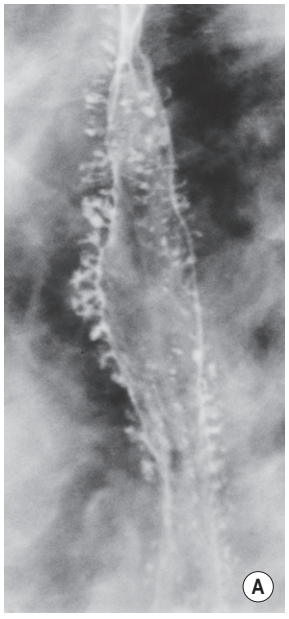
- The excretory ducts of the oesophageal deep mucous glands dilate and fill with barium ► they are seen on barium studies as multiple, flask-shaped mucosal outpouchings ► this disease is usually diffuse, but may be localized if it is associated with peptic stricture formation or an oesophageal carcinoma
- Fistulation may occur between these pseudodiverticula ► intramural abscesses may develop which can rarely perforate through the oesophageal wall ► long tapered strictures may arise
- It is associated with oesophagitis (usually due to reflux) ► other underlying disorders include diabetes, candidiasis and alcoholism

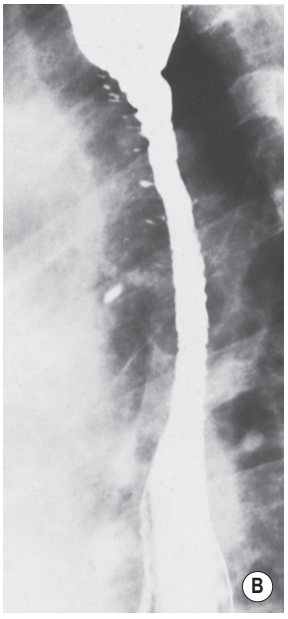




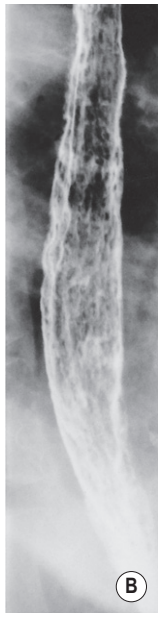
An annular peptic stricture at the GOJ. (A) An area gastricae pattern is present below the stricture. (B) Benign peptic stricture above a hiatus hernia. The stricture has smooth tapered margins. (C) Benign peptic stricture. Asymmetric ulceration and scarring has produced a stricture with irregular and shoulder margins resembling a carcinoma. Erosions on the oesophageal folds give them a lobular margin resembling varices (arrows).†

Corrosive stricture. A long stricture extending up to the mid-oesophagus (resulting from swallowing lye as child).†

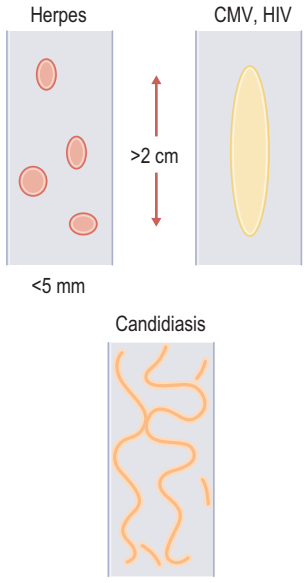




Intramural pseudodiverticulosis. (A) Multiple flask-shaped projections produced by barium entering dilated oesophageal glands. (B) Mid-oesophageal stricture with small flask-shaped projections.†



Candida oesophagitis (A) Mucosal plaques. (B) Extensive mucosal nodularity.†



Diagrammatic representation of oesophageal ulceration.

BENIGN TUMOURS

DEFINITION

- Benign tumours arising from the oesophageal mucosa or submucosa:
  - *Mucosal origin:* papilloma
  - *Submucosal origin:* leiomyoma (the commonest type) ► neurofibroma ► lipoma ► fibrovascular polyp

CLINICAL PRESENTATION

- These can be asymptomatic or present with dysphagia
  - *Fibrovascular polyp:* this may be regurgitated into the mouth and even, on occasion, aspirated (resulting in asphyxia)

RADIOLOGICAL FEATURES

Barium swallow

- **Papilloma:** these are usually small (2–5mm) ► larger papillomas may trap barium within the interlacing fronds that cover their surface
- **Leiomyoma:** these are usually found within the lower $^1/_3$ of the oesophagus ► they appear as a smooth wide-based filling defect covered by an intact mucosa ► they may calcify and can be multiple
- **Neurofibroma/lipoma:** these may be difficult to distinguish from a leiomyoma and are extremely rare
- **Fibrovascular polyp:** these are usually found within the proximal oesophagus ► they are pedunculated (the stalk forms due to repeated passage of food with peristalsis) ► they may expand the oesophageal lumen but rarely cause significant barium hold-up (due to their very pliable nature)

PEARLS

- **Glycogenic acanthosis:** this results from the accumulation of glycogen within the squamous epithelium (and is therefore not a tumour as such) ► its aetiology is unclear (but possibly age related) ► it demonstrates no malignant potential
  - *Endoscopy:* small white or yellow plaques measuring 2–5 mm in size

MALIGNANT TUMOURS

DEFINITION

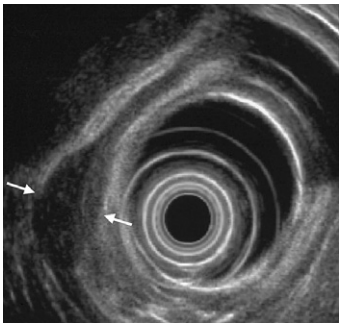
- Malignant tumours arising from the oesophageal mucosa or submucosa
  - **Oesophageal carcinoma**: the commonest malignant tumour (see separate section)
  - **Leiomyosarcoma** (1%): these arise from the *smooth* muscle within the oesophageal wall – therefore they are found only within the distal oesophagus (*striated* muscle is found within the proximal $^1/_3$ of the oesophagus) ► they can grow to an extraordinary size before symptoms present due to their failure to cause obstruction ► they are relatively indolent and metastasize late
  - **Melanoma** (1%): these are rare tumours (melanoblasts are uncommon within the oesophagus) ► they metastasize early with a very poor prognosis
  - **Lymphoma** (1%): oesophageal involvement is very rare ► it is usually of the non-Hodgkin's type and is usually associated with lymphomatous disease elsewhere
    - It begins as a submucosal lesion (usually in the distal $^1/_3$ of the oesophagus) resulting in a smooth luminal narrowing with an intact overlying mucosa ► later ulceration can develop
    - Secondary involvement by contiguous spread from adjacent nodal disease is more common but rarely results in dysphagia
  - **Spindle cell carcinoma**: a rare tumour containing both carcinomatous and spindle cell elements
  - **Metastases**: these are usually due to direct extension from tumours within the thoracic cavity (notably carcinoma of the bronchus) ► involved nodes may also infiltrate the oesophagus causing displacement and occasionally fistula formation between the oesophageal lumen and the adjacent bronchus ► carcinoma of the pancreas (particularly the tail) may involve the distal oesophagus or gastro-oesophageal junction
    - Breast carcinoma is the most common distant cause of oesophageal metastases

RADIOLOGICAL FEATURES

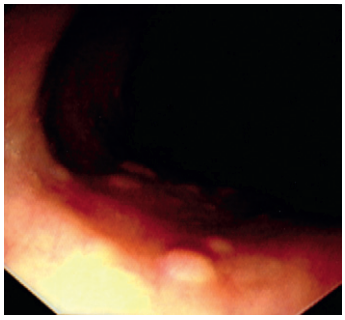
Barium swallow/CT

- **Leiomyosarcoma:** a large polypoid mass ► there can be a large exophytic component which may be seen on a CXR as a mediastinal mass
- **Melanoma:** a large polypoid mass (which will appear black on endoscopy)
- **Lymphoma:** this commonly involves the distal oesophagus following spread of lymphoma from the stomach ► the oesophagus may show widespread changes due to submucosal infiltration (presenting as multiple nodules)
- **Spindle cell carcinoma:** a bulky, polypoid tumour within the mid-oesophagus
- **Metastases:** involvement of the distal oesophagus and GOJ by carcinoma of the pancreas may lead to a right-angled bend of the distal oesophagus
  - Breast carcinoma usually causes submucosal masses

Leiomyoma of the oesophagus. Two views showing features typical of an intramural or extrinsic lesion. There is a broad-based filling defect bulging into, and widening, the lumen of the oesophagus.†



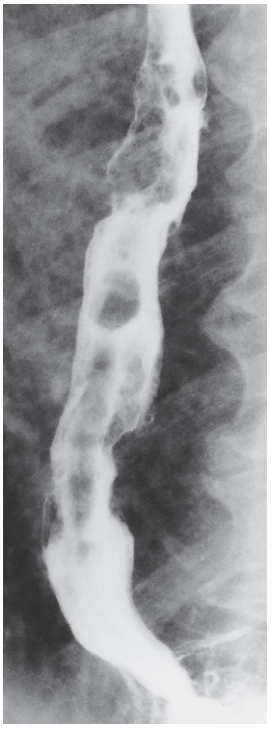
Leiomyoma. Endoscopic ultrasound demonstrating an echo-poor mass (between arrows) that is continuous with the layer of the muscularis propria.†



An endoscopic view of glycogenic acanthosis showing the typical small, smooth plaques of this condition.*

Spindle cell sarcoma. A bulky polypoid tumour arising in the mid-oesophagus.†

Barium study of a stricturing adenocarcinoma situated just above the gastro-oesophageal junction, as demonstrated by irregularity and an abrupt transition to normal mucosa on its cranial side.*



Oesophageal carcinoma with submucosal extension simulating varices, however (unlike varices) the width of the elongated filling defect was not influenced by the degree of oesophageal distension.

OESOPHAGEAL CARCINOMA

DEFINITION

- A malignant tumour of the oesophagus:
  - *Squamous cell carcinoma (>70%):* this arises from the squamous epithelium ► it is commonly found within the mid-oesophagus (50%), but rarely invades the stomach (cf. an adenocarcinoma)
    - It is associated with smoking, alcohol and tobacco consumption
    - Less common associations include: achalasia ► Plummer–Vinson syndrome ► head and neck tumours ► coeliac disease ► radiation change
  - *Adenocarcinoma (<30%):* this is due to malignant degeneration of columnar metaplasia ► the majority (90%) are found within the distal $^1/_3$ of the oesophagus ► there is an increasing frequency relative to squamous carcinoma due to an increasing prevalence of Barrett's oesophagus and scleroderma (the patulous oesophagus leads to reflux)

CLINICAL PRESENTATION

- It is rare under the age of 40, with an increasing incidence with age thereafter (3M:1F)
  - It is often advanced with a poor prognosis at presentation: the lack of oesophageal serosa does not provide a barrier to tumour spread ► there are multiple mediastinal structures that are close by
- *Presenting features:* dysphagia (indicating unresectable disease) ► weight loss ► recurrent aspiration ► early satiety (with distal adenocarcinomas) ► fistula formation with the tracheobronchial tree (seen in up to 10% of squamous cell carcinomas)

RADIOLOGICAL FEATURES

- There are a wide range of morphological appearances: nodular, polypoid, ulcerative, or irregular structuring
  - There can also be an uncommon varicoid (or superficial spreading) type

Barium swallow

- **Early:** stiffened mucosa ► a failure to collapse completely following a peristaltic wave
- **Late:** an irregular stricture with nodular or rolled margins ► the ulcerative type usually has a firm irregular tumour rim
- **Varicoid carcinoma:** this follows submucosal spread of tumour ► it results in tortuous thickened folds that can mimic varices
- **GOJ tumours:** an achalasia-like picture due to infiltration and destruction of the myenteric plexus ► nodules and irregular margins can be seen at the GOJ
- **Advanced cases:** fistulation with the tracheobronchial tree (leading to recurrent aspiration) ► total dysphagia

CT

- Asymmetric oesophageal wall thickening ($\geq$ 5mm) ► a dilated oesophagus cranial to the tumour (due to obstruction)
- The tumour may extend through the serosa into the peri-oesophageal fat with loss of the sharp interface, together with the formation of a high attenuation mass ► it may invade any adjacent structures (e.g. the aorta)
- Rarer polypoid tumours are shown as intraluminal filling defects that may expand the overall oesophageal diameter
- CT is excellent at assessing local gastro-hepatic nodes ► however it is poor at assessing the extent of tumour invasion of the oesophageal wall (i.e. differentiating between T1 and T2 tumours)
  - Ill-defined para-oesophageal fat suggests T3 disease

MRI

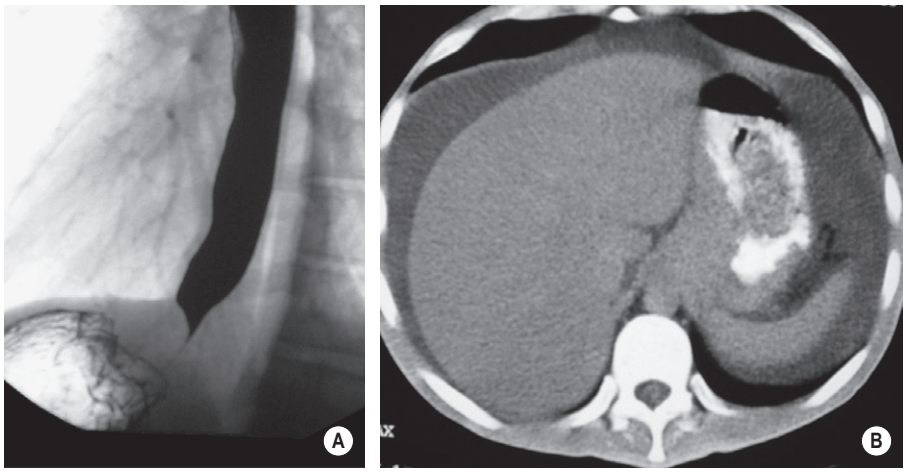
- Like CT, it is also unable to distinguish between T1 and T2 disease
- T1WI: tumour is isointense to the normal oesophagus (extra-oesophageal invasion is suggested by loss of the high SI from the adjacent fat)
- T2WI: intermediate to high SI tumour mass
- T1WI + Gad: enhancement

EUS

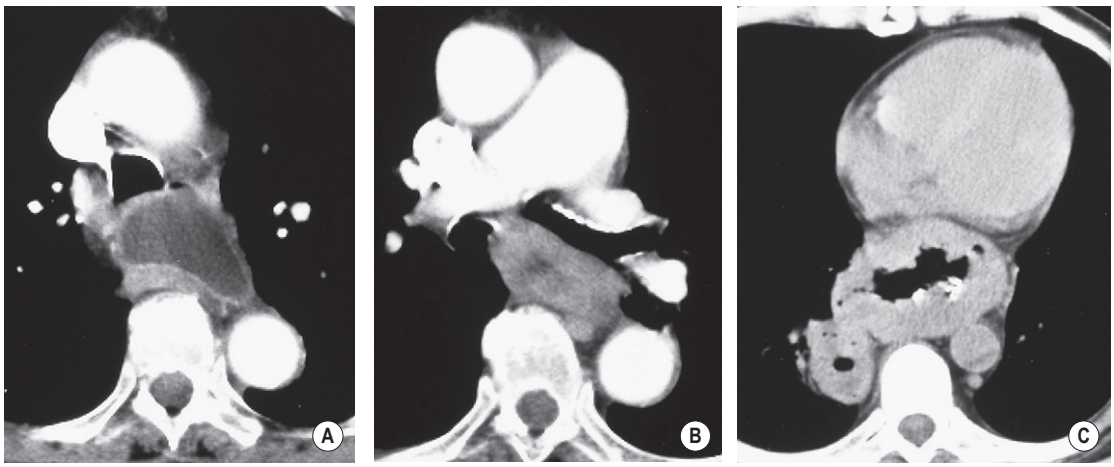
- A hypoechoic tumour disrupting the normal 5 oesophageal wall layers
  - This is the only modality that can currently differentiate between a T1 (just involving the mucosa) and a T2 (invading the muscularis propria) tumour ► a T3 lesion will demonstrate adventitial infiltration
- It can identify malignant local nodes: these are well marginated, spherical, hypoechoic, and demonstrate loss of their internal structure
  - Elongated nodes are more likely to be benign
- *Limitations:* it cannot pass a tight stricture ► it may overestimate the tumour extent (due to peritumoral inflammation) ► it may not be able to distinguish reactive from metastatic lymph nodes

FDG PET and FDG PET-CT

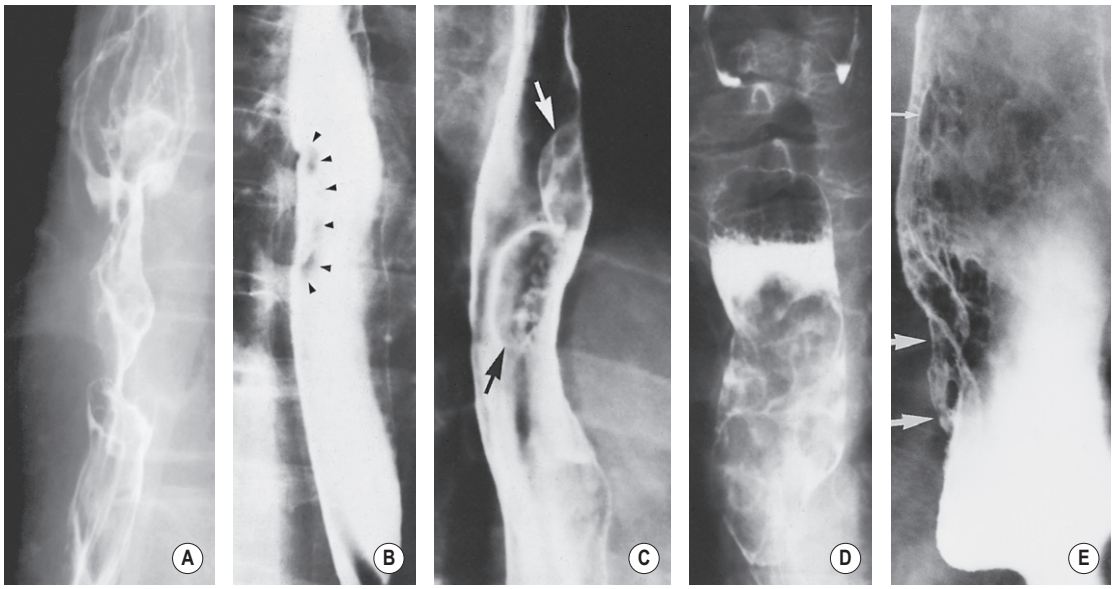
- Oesophageal carcinomas are FDG avid
- This is more sensitive, specific and accurate than CT in identifying loco-regional lymphadenopathy and thus the overall staging ► PET-CT is more sensitive than PET alone
- It can define the craniocaudal tumour extent and also can assess any chemotherapeutic response (particularly the differentiation of malignant disease from residual oesophageal thickening)



Pseudo achalasia appearance. Note the tapered appearance of the distal oesophagus which superficially resembles an achalasia, but the clue to the sinister nature of this lesion is from the impression on the gastric fundus.†



Typical CT appearances of oesophageal carcinoma. Note the dilated oesophagus (A) situated cranial to an area of wall thickening (B). (C) CT showing gross thickening of the oesophageal wall due to an extensive carcinoma that has also extended into the right lung base, producing an associated thick-walled cavity.*



Oesophageal carcinoma. (A) An advanced stricturing lesion with mucosal destruction and 'shouldering'. (B) A shallow ulcer with a tumour ring. (C) Two tumour nodules. (D) An irregular polypoidal intraluminal filling defect which is causing obstruction. (E) Early carcinoma. A plaque-like squamous carcinoma (large arrows) with a small satellite lesion caused by lymphatic spread (small arrow).

OESOPHAGEAL CARCINOMA

PEARLS

Normal oesophageal wall layers on Endoscopic Ultrasound (EUS)

- Superficial mucosa: *hyperechoic*
- Muscularis mucosae: *hypoechoic*
- Submucosa: *hyperechoic*
- Muscularis propria: *hypoechoic*
- Adventitia: *hyperechoic*

Mechanisms of tumour spread

- ***Lymphatic spread:*** nodal involvement occurs at the same level as the tumour (± paratracheal, hilar and para-aortic adenopathy)
  - Cranial longitudinal spread can occur to the internal jugular, cervical and supraclavicular nodes
  - Caudal longitudinal spread can occur to the left gastric and coeliac nodes
- ***Haematogenous spread:*** this occurs early and is commonly to the lungs and liver (and less commonly to bone)
- ***Direct invasion***
  - *Aortic invasion:* surgical resectability of the primary tumour is less likely to be successful if there is $\geq 90^{\circ}$ of contact with the aortic circumference, or if there is obliteration of the small triangle of fat between the oesophagus, aorta and spine
  - *Tracheobronchial invasion:* there is inward bowing of the posterior tracheal wall (which normally has a convex appearance) ► there is tracheal or bronchial displacement away from the spine ► there is obliteration of the fat plane between the oesophagus and the upper respiratory tract

Treatment and palliation of oesophageal cancer

- ***Treatment:*** primary surgical resection if the tumour involves the mucosa or submucosa ► neoadjuvant therapy if there is extension into the muscularis propria (or beyond)
- ***Palliation for advanced disease:*** stents are used when there is progressive dysphagia with an inability to swallow saliva
  - *Stent delivery:* either at endoscopy (via the endoscope biopsy channel) or under direct radiological control
  - *Self-expanding metal stents:* these are delivered on a small flexible introducer ► they are made of stainless steel or nitinol
    - Nitinol: a compound of nickel and titanium which has an inbuilt memory feature allowing it to return to a preset size and shape following delivery
  - *Covered stents* (coating of polyurethane): these are useful to reduce tumour overgrowth and for dealing with fistulae ► they are unsuitable for deployment across the GOJ (as there can be significant migration)
  - *Complications:* oesophageal perforation ► distal stent migration into the stomach ► gastro-oesophageal reflux (if a stent is placed across the GOJ) ► possible tracheal compression or a persistent sensation in the throat (with high stent placement)

BARRETT'S OESOPHAGUS

DEFINITION

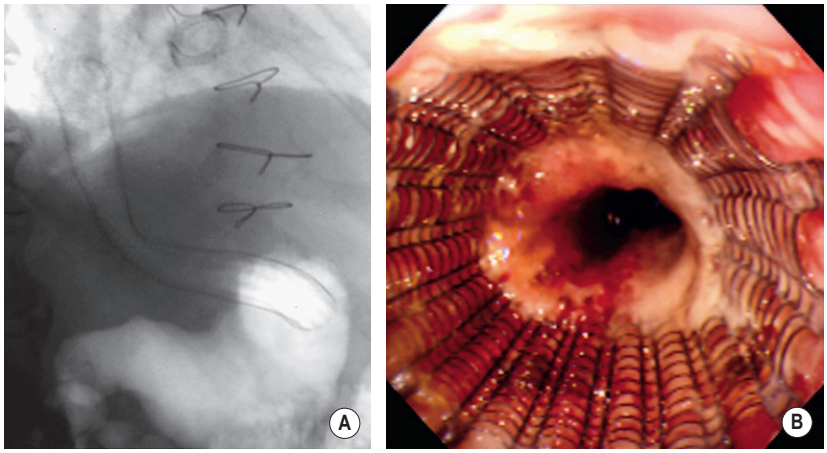
- Chronic gastro-oesophageal reflux causes a specialized non-secretory columnar epithelium to grow cranially into areas previously covered by squamous epithelium
  - Usually 2cm or more of columnar epithelium is required before the term is used

RADIOLOGICAL FEATURES

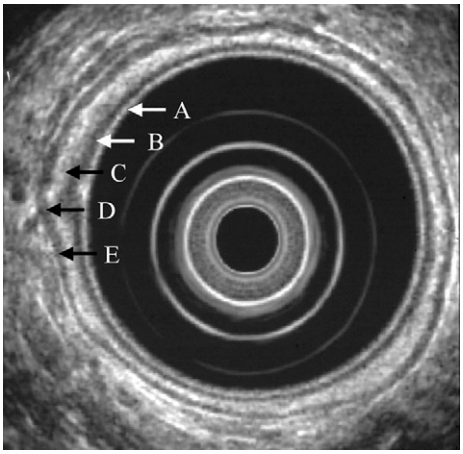
- **Barium swallow:** there is a wide patulous hiatal segment (associated with a hiatus hernia) ► there is a dilated segment of oesophagus above the hernia (lined by columnar epithelium) which often appears to be 'bell' or 'tent' shaped
  - The junction of the two mucosal types is marked by a ring (which is often slightly contractile) where ulceration and secondary strictures may form ► this may be some way above the hiatus (at the level of the aortic knuckle or above)

PEARLS

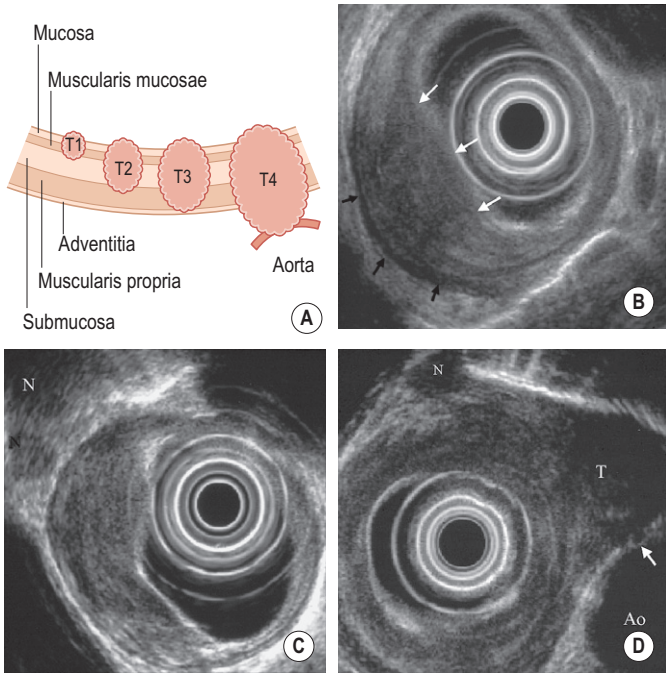
- It is ultimately diagnosed with endoscopy and biopsy
- Barrett's oesophagus is associated with up to a 40-fold increased risk of developing oesophageal carcinoma (15% of patients with a Barrett's oesophagus develop an adenocarcinoma) ► the precursor is a high-grade dysplasia and therefore regular endoscopic screening is required



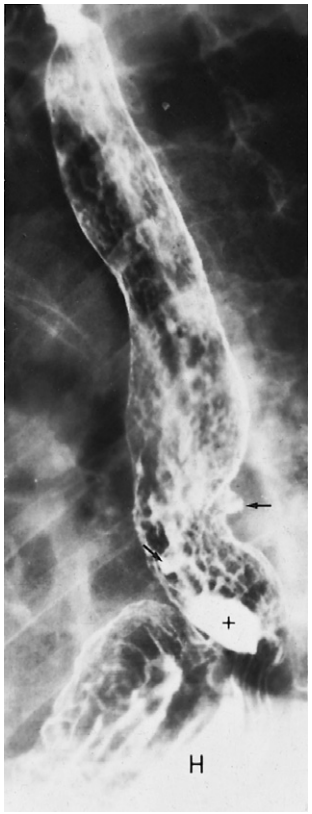
(A) An oesophageal stent in situ across a junctional tumour. (B) Endoscopic view of an oesophageal stent in position. Note the tumour ingrowth through the mesh of the distal part of this stent.*



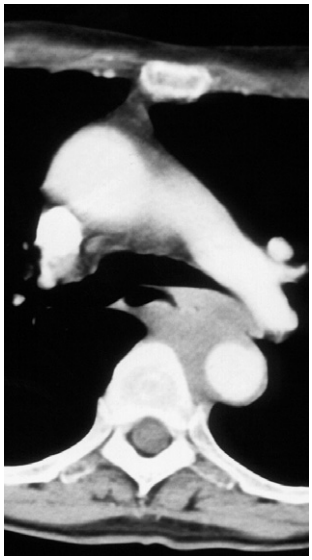
EUS of the oesophagus showing the wall layers. A = mucosa, B = muscularis mucosae, C = submucosa, D = muscularis propria, E = adventitia.†



EUS staging of oesophageal carcinoma. (A) T staging: T1 (limited to the mucosa and submucosa), T2 (involves the muscularis propria), T3 (involves the adventitia), T4 (invades adjacent structures). (B) T2 tumour – the muscularis propria has not been breached (black arrows). The luminal tumour border is indicated by the white arrows. (C) T2N1 tumour. The muscularis propria has not been breached but there are adjacent involved lymph nodes (N). Malignant nodes are round, hypoechoic and well defined with a loss of their internal structure. (D) T4 – tumour has breached the muscularis propria and is invading the aortic wall (arrow). Ao = aorta, T = tumour, N = metastatic lymph node.†



Barrett's oesophagus. Ulceration (arrows) at the squamocolumnar junction, below which is a fine reticular pattern. This resembles the areae gastricae pattern of the stomach, and is produced by islands of columnar mucosa. + = pool of barium, H = hiatus hernia.†



CT appearances of a mid-oesophageal carcinoma, which demonstrates extensive soft tissue involvement around the adjacent aorta, thus rendering it inoperable.*

ACHALASIA

DEFINITION

- A motor disorder of the oesophagus caused by the degeneration of the neurones of Auerbach's plexus (which is situated between the oesophageal longitudinal and circular muscle coats)
  - This leads to a failure of relaxation of the GOJ

CLINICAL PRESENTATION

- The onset occurs between 20 and 40 years of age
- It presents initially with dysphagia for both solids and liquids (cf. strictures which initially only cause dysphagia for solids)
  - There is a recurrent aspiration and pneumonia risk (10%)

RADIOLOGICAL FEATURES

Barium swallow

- ***Early:*** defective distal peristalsis is associated with a slight narrowing at the GOJ
  - *'Vigorous achalasia':* marked non-propulsive contractions may occur
- ***Late:*** the body of the oesophagus becomes progressively dilated and aperistaltic with a subsequent huge oesophageal residue of food and fluid debris (this can be seen as a fluid level on a CXR and is a significant aspiration risk)
  - *'Rat's tail' or 'bird's beak' appearance:* a characteristic appearance of the gastro-oesophageal junction

PEARLS

- Oesophageal squamous cell carcinoma is a likely complication with severe long-standing (> 20 years) disease
- In patients with early achalasia, drinking hot water in the erect position during fluoroscopy produces an immediate and pronounced relaxation of the GOJ
  - The diagnosis still needs to be confirmed by manometry (± scintigraphy) ► endoscopy will exclude a carcinoma as a cause of secondary achalasia

DIFFUSE OESOPHAGEAL SPASM

DEFINITION

- A dysmotility disorder of the oesophagus (it is five times less common than achalasia)
- It is characterized by strong repetitive non-propulsive contractions which may be interspersed with normal peristaltic waves ► these marked contractions may completely obliterate the oesophageal lumen

CLINICAL PRESENTATION

- Chest pain (it may or may not be associated with swallowing)

RADIOLOGICAL FEATURES

Barium swallow

- Strong non-propulsive contractions may lead to a 'corkscrew' or 'curling' oesophagus
- There may be marked oesophageal wall thickening (which may extend to a depth of up to several cm)
- Occasionally there can be oesophageal diverticula (as a consequence of the high pressure contractions)

OTHER CAUSES OF DYSMOTILITY

GORD This is probably the commonest cause of oesophageal dysmotility

Presbyoesophagus An age-related (>70 years) oesophageal dysmotility which is not attributable to a specific condition

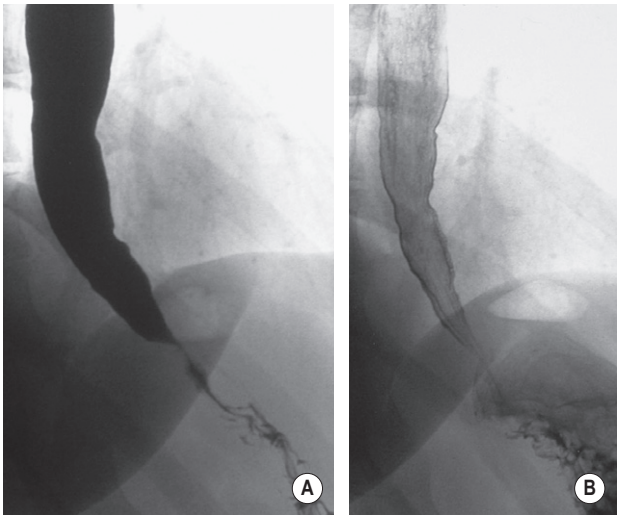
Scleroderma This is the commonest systemic disease to cause oesophageal dysmotility ► it is a collagen vascular disorder of unknown aetiology characterized by smooth muscle atrophy followed by collagen deposition and fibrosis involving the oesophagus, stomach and small bowel

Barium swallow Initially there is diminished peristalsis (or tertiary contractions in the distal ⅓ of oesophagus) – this is then followed by absent peristalsis ► the GOJ becomes wide and patulous resulting in GOR, reflux oesophagitis and stricture formation ► oesophageal stasis may result in *Candida* colonization and an increased risk of adenocarcinoma

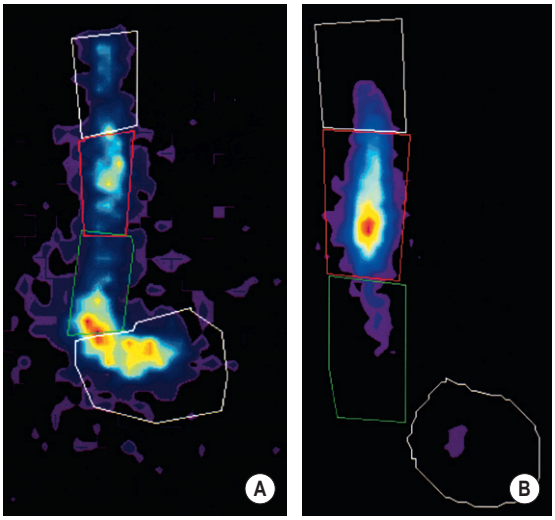
Chagas disease This is caused by *Trypanosoma cruzi* which produces a neurotoxin destroying the ganglion cells within the myenteric plexus ► it can affect the oesophagus, colon (megacolon or sigmoid volvulus), stomach (megastomach), duodenum (megaduodenum), heart (cardiomyopathy) and CNS (encephalitis)

Barium swallow Initially the oesophagus will demonstrate hypercontractility and distal muscular spasm ► during the later stages an appearance similar to achalasia can present when denervation has occurred

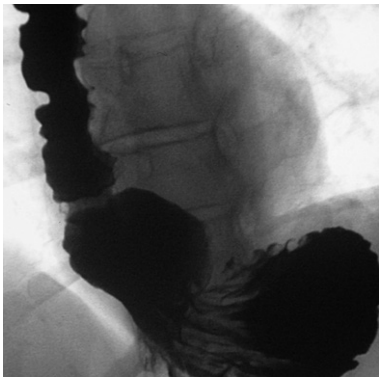
- Oesophageal complications include ulceration, perforation and carcinoma



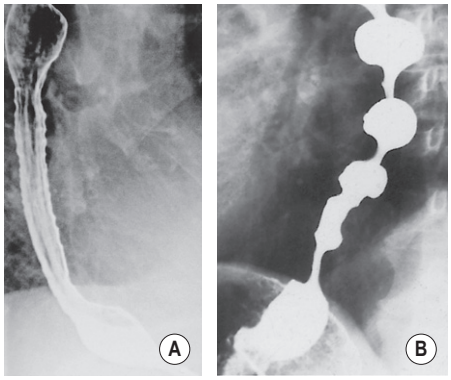
Classical appearances of achalasia. Note the tapered appearance of the GOJ with the column of barium above (A). As soon as hot water has been drunk, the whole barium column falls through into the stomach (B).*



Radio-isotope study also demonstrating achalasia. A normal examination is shown with isotope in the stomach (A), whereas in achalasia the isotope is retained in the oesophagus (B).*



View of a large sliding hiatal hernia that demonstrates gross spontaneous gastro-oesophageal reflux when the patient lifts the left side while in the supine position. Note also the marked oesophageal incoordination produced by the reflux.*



Tertiary contractions of the oesophagus seen as (A) a rippling of the oesophageal wall or (B) a series of indentations resembling a corkscrew (hence the description 'corkscrew oesophagus').†

Scleroderma. Incompetence of the gastro-oesophageal sphincter resulting in severe reflux oesophagitis with stricturing, oedematous mucosa (mosaic pattern) and deep ulceration.†

Differentiating scleroderma from achalasia

	Scleroderma	Achalasia
Oesophageal dilatation	Mild	Significant
GOJ	Wide and patulous	Characteristic 'rat's tail' or 'bird's beak' appearance
Complications	Reflux ± late stricture formation ± Barrett's oesophagus	Aspiration

OESOPHAGEAL ATRESIA (OA) AND TRACHEO-OESOPHAGEAL FISTULA (TOF)

DEFINITION

- This is due to abnormal partitioning of the laryngotracheal tube from the oesophagus by the tracheo-oesophageal septum during the 4th week of gestation ► it affects 1:3000–4500 live births
  - The atretic segment of the oesophagus tends to be at the junction of its proximal and middle thirds
  - A TOF (if present) is usually found proximal to the carina
  - Occasionally an isolated TOF can occur without an oesophageal atresia (the 'H'- or 'N'-type fistula)
- 50% of patients have associated congenital anomalies:
  - *VACTERL spectrum:* **V**ertebral anomalies ► an **A**norectal malformation ► **C**ardiovascular malformations (VSD, PDA, right aortic arch) ► **T**racheal anomalies ► an o**E**sophageal fistula ► **R**enal anomalies ► **L**imb anomalies
  - *Other anomalies:* duodenal atresia and stenosis ► an imperforate anus ► trisomy 18 and 21 ► Potter's syndrome

CLINICAL PRESENTATION

- **Antenatal (US):** maternal polyhydramnios
- **Postnatal:** an immediate presentation with choking, coughing, cyanosis and drooling (this is exacerbated during attempts to feed)
  - H-type fistulas generally present later in infancy or childhood with episodes of choking or apnoea during feeding, or recurrent respiratory tract infections

RADIOLOGICAL FEATURES

CXR An orogastric tube will curl up within the proximal oesophageal pouch (there can also be aspiration pneumonitis, vertebral anomalies, or an abnormal cardiac silhouette)

- Gas within the abdomen implies a distal fistula (neonates with a H-type fistula commonly have a abdomen distended with gas)
- A gasless abdomen implies an isolated oesophageal atresia (or an atresia with a proximal fistula) ► there is an absent fetal gastric bubble if there is an oesophageal atresia (but no TOF)
- *Isolated oesophageal atresia:* a long gap between the atretic segments is seen in association with 13 pairs of ribs

Upper GI study This can delineate a H-type fistula: the patient is placed prone and a horizontal X-ray is used ► contrast medium is injected under pressure (via a nasogastric tube with its tip in the distal oesophagus) and the tube is then slowly withdrawn under fluoroscopic guidance

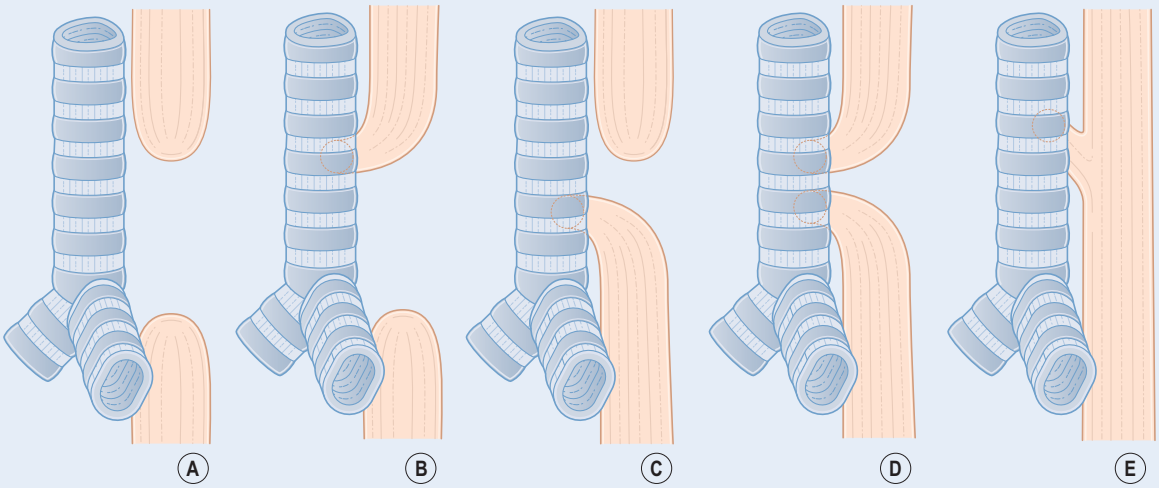
- The majority of these fistulas are seen at the level of the thoracic inlet
- *The gap between the oesophageal pouches can be assessed following the formation of a feeding gastrostomy:* under fluoroscopic guidance, a Heger dilator is inserted through the gastrostomy and retrogradely into the distal oesophagus ► a Repogle tube is simultaneously used to delineate the superior pouch ► as both tubes are radio-opaque, the degree of separation can be easily assessed
  - Alternatively CT can delineate the gap following the simultaneous injection of air into the upper pouch (via the Repogle tube) and via the gastrostomy

PEARLS

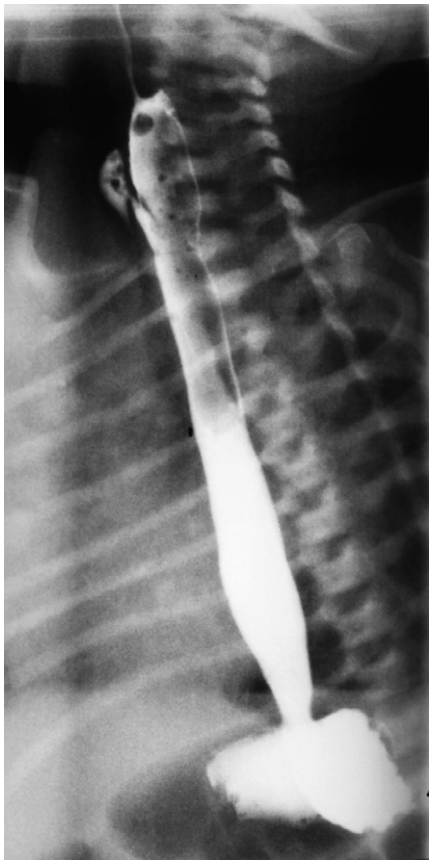
- The mortality rates are now no longer due to the oesophageal atresia itself, but due to the associated malformations
- A combined bronchoscopy and oesophagoscopy should be performed if there is a high clinical index of suspicion of a H-type fistula with negative imaging

Complications following surgical repair

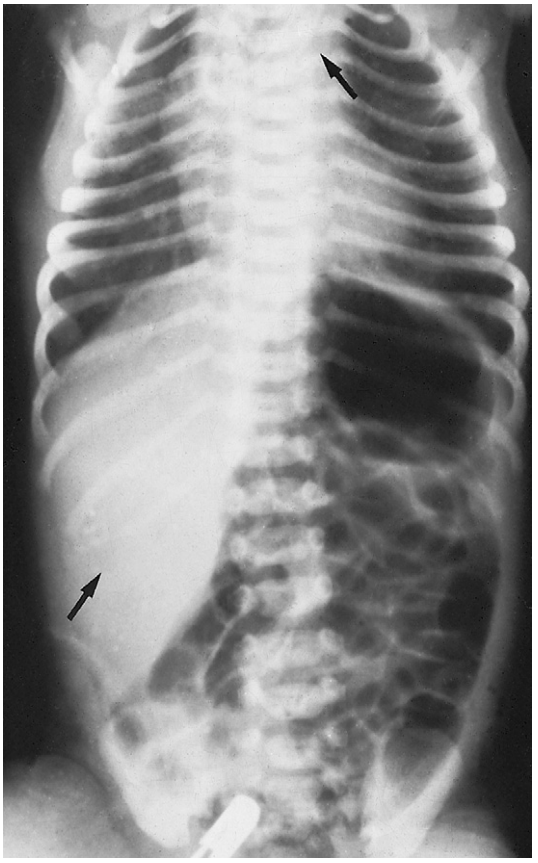
- *Recurrent TOF (10%):* this should be suspected if the oesophagus is gas filled on CXR and if contrast medium studies show 'beaking' of the anterior oesophageal wall
- *Other complications:* anastomotic breakdown (10–20%) ► anastomotic strictures ► disordered oesophageal and distal GI motility ► gastro-oesophageal reflux



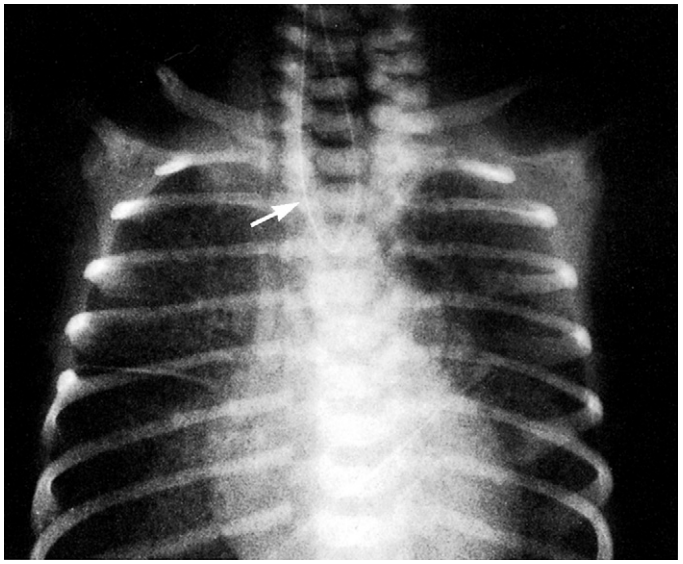
OA and TOF classification: Type (A) Isolated OA (8%) ► Type (B) Proximal fistula with distal OA (1%) ► Type (C) Proximal OA with distal fistula (85%) ► Type (D) Double fistula with intervening OA (1%) ► Type (E) Isolated fistula (H type) with no OA (4%).



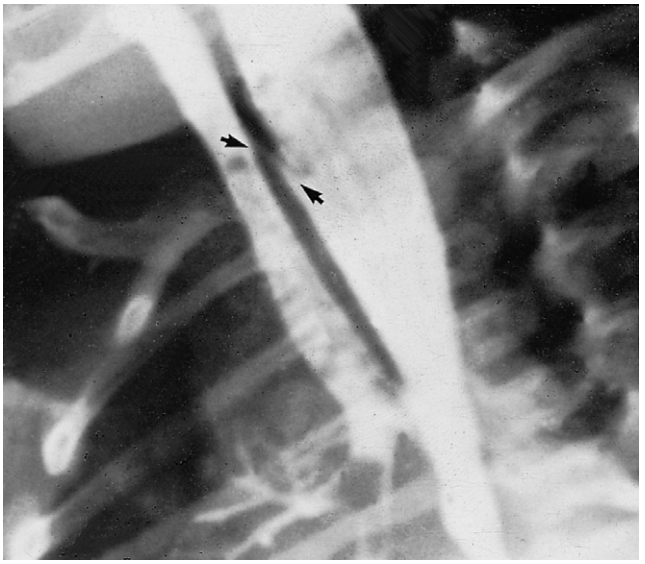
H-type TOF. Upper GI contrast study shows the fistula running obliquely at the level of the thoracic inlet.*



OA with TOF. A coiled nasogastric tube is seen in the dilated proximal oesophageal pouch (top arrow). The presence of distal air-filled bowel implies an associated TOF. Thirteen pairs of ribs are noted, compatible with VATER syndrome (lower arrow).



Oesophageal atresia. Supine CXR shows orogastric tube (arrow) curled in the proximal oesophageal pouch.*



Contrast oesophagram demonstrating oblique track (arrows) of a TOF with contrast filling the tracheobronchial tree.†

OESOPHAGEAL DIVERTICULA

Pulsion diverticulum This results from high intraluminal pressures acting on a potential oesophageal wall weakness

- ***Zenker's diverticulum (pharyngeal pouch):*** a false diverticulum originating within the posterior midline of the hypopharynx and above cricopharyngeus (Killian's dehiscence) ► it presents with dysphagia, retention of food debris within the diverticulum, halitosis and possibly a neck mass
  - *Complications:* ulceration ► aspiration pneumonia ► carcinoma
- ***Killian–Jamieson diverticulum:*** this is located off the midline and below cricopharyngeus (arising from the lateral cervical oesophageal wall) ► it is smaller and less symptomatic than a Zenker's diverticulum
- ***Epiphrenic diverticulum:*** this is located within the distal oesophagus and is the result of oesophageal contractions against a closed lower oesophageal sphincter
- ***Intramural pseudodiverticulosis:*** the dilated mucous glands within the oesophageal wall can simulate true diverticula on contrast studies

Traction diverticulum These result from an adjacent external inflammatory process generating pulling forces on the oesophageal wall (e.g. inflamed adjacent lymph nodes)

VARICES

Definition Dilated oesophageal submucosal veins act as a collateral venous drainage pathway in the presence of an obstruction elsewhere ► it is very rarely idiopathic

- **Uphill varices:** these affect the lower oesophagus ► they are caused by cirrhosis of the liver – portal blood flow is rerouted (via the oesophagus) to the SVC
- **Downhill varices:** these affect the upper $^{2}/_{3}$ of the oesophagus ► SVC obstruction causes blood to be rerouted from the head and neck (via the oesophagus) into the azygos vein ► if the azygos vein is also blocked then downhill varices may also be found within the lower oesophagus (taking blood to the coronary and portal vein)

Barium swallow Characteristic serpiginous filling defects best appreciated on the prone swallow examination (after administration of an IV anti-peristaltic agent)

CT Enhancing vascular venous structures around the oesophagus

OESOPHAGEAL WEBS

Definition These are common shelf-like (1–2mm thick) mucosal infoldings protruding into the oesophageal lumen ► they can be semicircular or form a complete ring ► they are usually found at the anterior aspect of the proximal cervical oesophagus

- They are usually asymptomatic but may cause dysphagia

Barium swallow A thin transverse filling defect (which may be circumferential)

Pearl As well as being spontaneous, they can be associated with: pemphigoid ► epidermolysis bullosa ► graft-versus-host disease ► reflux oesophagitis (a web within the distal oesophagus) ► Plummer–Vinson syndrome (a web + iron deficiency anaemia + dysphagia)

EXTRINSIC LESIONS

- **Thyroid masses**
- **Lymph node masses:** these are usually secondary to a bronchial carcinoma or lymphoma, affecting both the superior and mid-oesophagus
- **Fibrotic changes at the lung apices:** these can pull the oesophagus sideways, which may then form an acute angle within the superior mediastinum
- **Carcinoma of the bronchus**
- **Vascular causes**
  - *Aberrant right subclavian artery*: this gives rise to the classic appearance of a posterior impression on the proximal oesophagus or a band-like impression as the vessel ascends posterior to the oesophagus (AP view)
  - *A right-sided aortic arch:* an impression on the right side of the oesophagus
  - *Descending thoracic aortic aneurysm:* the aorta may erode into the oesophagus with catastrophic haematemesis
  - *An enlarged left atrium:* this displaces the distal $^{1}/_{3}$ of the oesophagus
- **Cricopharyngeal spasm**
  - The cricopharyngeus normally relaxes during swallowing – if it remains contracted it forms a smooth posterior impression at the pharyngo-oesophageal junction (C5/6 level) ► this may cause dsyphagia and lead to a Zenker's diverticulum

TRAUMA

Definition This is usually iatrogenic and commonly due to an oesophageal perforation following an endoscopy (due to an unsuspected pharyngeal pouch) ► other causes:

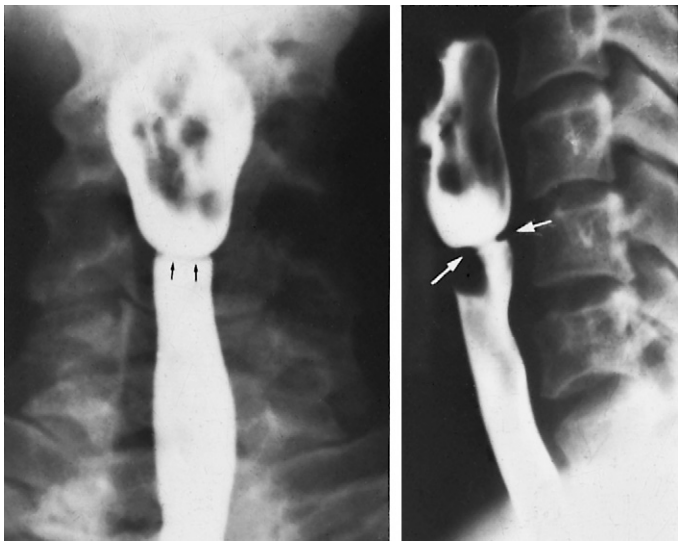
- *An ingested foreign body:* this can also cause an intramural haematoma or an oesophageal dissection
- *Severe vomiting affecting the distal oesophagus or proximal stomach:*
  - *Mallory–Weiss tear:* a mucosal tear where the wall integrity is maintained (with no pneumomediastinum)
  - *Boerhaave's syndrome:* a full-thickness tear (usually on the left posterolateral side) presenting with severe chest and epigastric pain ► immediate surgery is required

Radiological features of a full-thickness tear

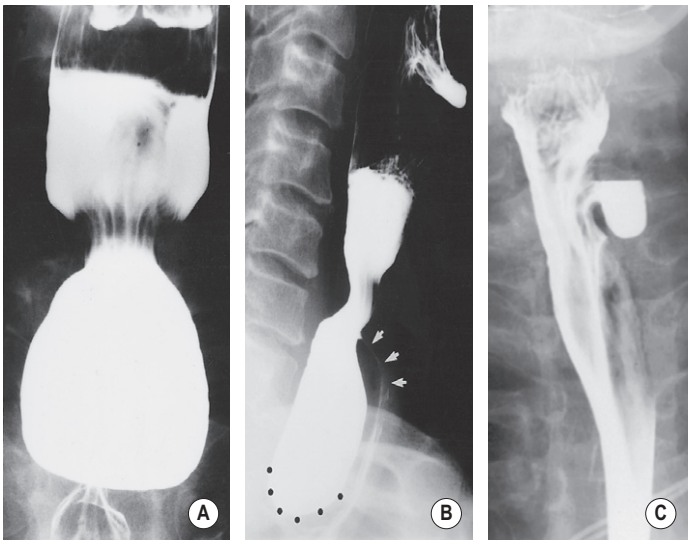
CXR Surgical emphysema within the neck ► pneumomediastinum ► a left pleural effusion

CT This can detect free gas adjacent to the oesophagus

Contrast swallow A leak is best demonstrated with a decubitus film (a horizontal X-ray beam)

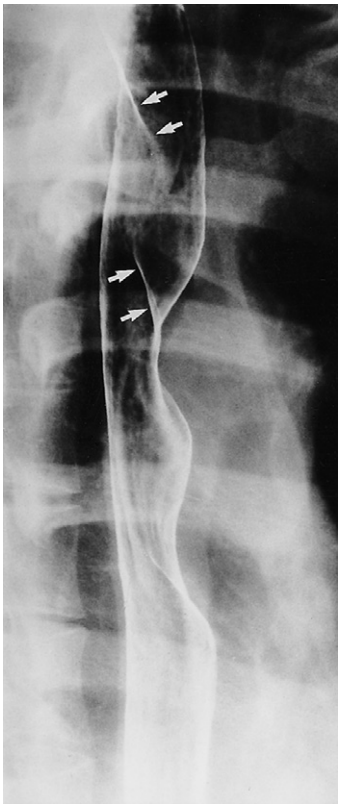


A concentric upper oesophageal web seen in both the frontal and lateral projections (arrows). The way in which the web narrows the lumen is well seen in the lateral view.†

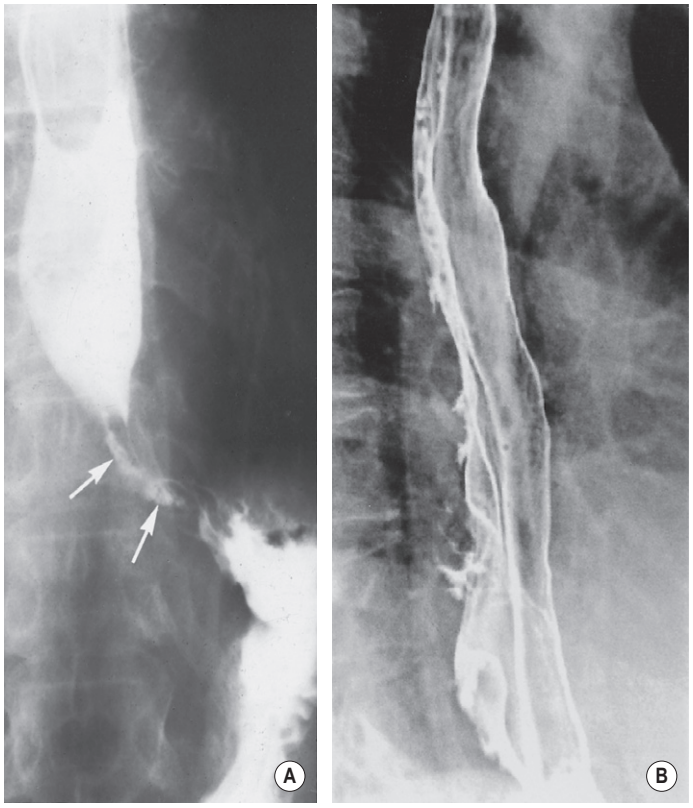


Zenker's pharyngeal diverticulum. (A) Frontal view. (B) Lateral view. Barium fills the diverticulum and then spills over into the anteriorly displaced oesophagus (arrows). (C) Lateral cervical oesophageal diverticulum (Killian–Jamieson diverticulum).†

Barium swallow showing the cervical oesophagus. Lateral view. The posterior impression (arrow) is produced by failure of the cricopharyngeus muscle to relax.†



Aberrant right subclavian artery producing characteristic extrinsic defect (arrows) of the oesophagus just above the level of the aortic arch. Left anterior oblique view.†



(A) Mallory–Weiss syndrome. Tear in the oesophagus at its lower end caused by vomiting. Barium (arrows) has tracked through the defect to lie beneath the mucosa. (B) Mucosal tear and intramural haematoma spreading along the length of one side of the oesophagus. The patient had swallowed a meat bone.†

3.2 STOMACH

BENIGN GASTRIC ULCERS

DEFINITION

- Gastric ulcers penetrate the stomach wall through the mucosa and into the submucosa (and frequently also into the muscularis propria) ► 95% are benign and they can also be multiple

CAUSES

- *Helicobacter pylori* infection (70%) ► NSAIDs ► alcohol abuse ► steroid use ► emotional stress ► smoking ► hereditary factors

RADIOLOGICAL FEATURES

Location Distal stomach > proximal stomach ► lesser > greater curvature ► posterior wall > anterior wall ► benign ulcers are rarely seen within the fundus

- *Ulcers due to NSAIDs*: these often affect the distal greater curvature (particularly the antrum) ► they are often multiple
- *Ulcers in the elderly*: these are more evenly distributed throughout the stomach (particularly affecting the proximal lesser curve)

Double-contrast barium studies (en face)

- The primary sign is a collection of barium on the dependent wall ► benign ulcers are usually round, oval or linear in shape ► a ring shadow may be seen if the ulcer is located on a non-dependent surface (as barium coats the edge of the ulcer crater)
- A smooth mound of surrounding oedema is seen as a circular filling defect ► the presence of normal areae gastricae extending to the ulcer crater is a good sign of a benign lesion

Double-contrast barium studies (in profile)

- A benign ulcer will project beyond the stomach lumen
  - *Hampton's line:* a thin lucent line crossing the ulcer base (representing preserved gastric mucosa with undermining of the more vulnerable submucosa) ► although rarely seen, this is virtually diagnostic of a benign ulcer
  - *Ulcer collar:* more commonly there is a thicker smooth rim of lucency at the ulcer base
  - *Ulcer mound:* with increasing oedema a symmetrical gently sloping mass can be seen

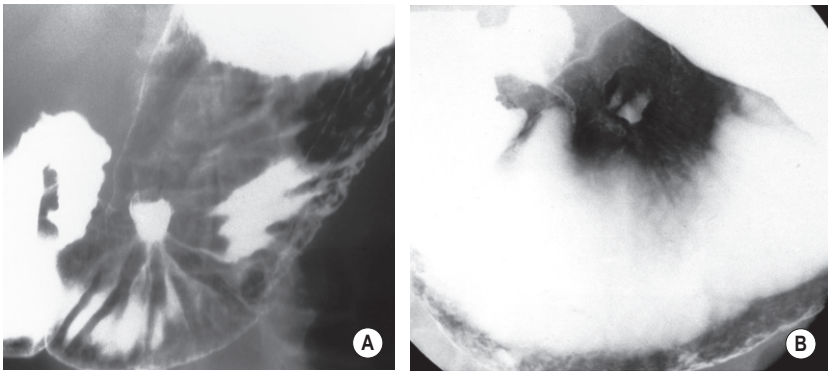
Benign vs malignant ulceration••

	Benign	Malignant
Location	Usually the antrum (75% on the lesser curve)	Usually the antrum (but can occur anywhere)
Fold convergence	To the crater edge	Stops short of the crater edge
Fold shape	Normal or uniformly swollen	Amputated, fused, or clubbed folds
Projection beyond gastric wall	Yes	No
Ulcer collar	Well defined	If present, it is irregular
Multiplicity	Common	Uncommon
Carman meniscus sign	No	Yes
Hampton's line	Yes	No
Response to therapy	Yes	No

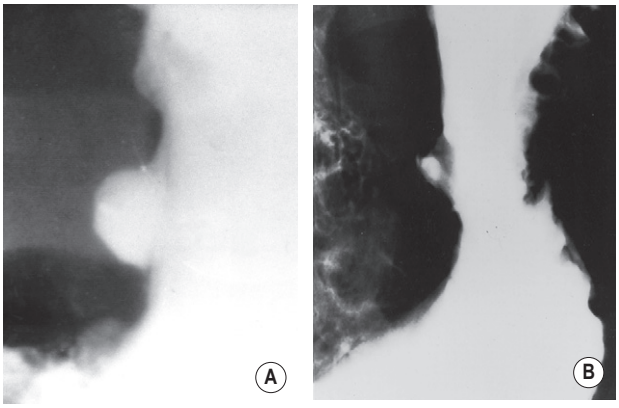
PEARLS

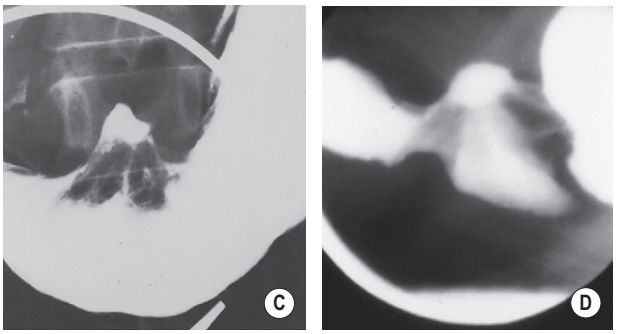
- **'Giant' ulcers (>3cm):** these are almost always benign but have a higher rate of complications (e.g. bleeding or perforation)
- **Healing ulcers:** > 95% heal within 8 weeks of medical treatment
  - As benign ulcers heal they may change shape from round or oval to linear crevices ► there may be a residual central pit or depression
  - Radiating folds seen with healing ulcers should be smooth, thin, symmetric and continue to the edge of the crater
  - There may be subtle retraction or wall stiffening ► a healed antral ulcer may form prominent transverse folds or significant antral narrowing with the deformity leading to obstruction
- **Features suspicious for malignancy:** location within the fundus or proximal ½ of the greater curvature ► incomplete healing ► irregularity of the radiating folds ► a residual mass ► loss of the mucosal pattern
- **Gastric erosions (aphthous ulcer):** these are shallow ulcerations that do not penetrate the muscularis mucosa ► they are usually seen with *H. pylori* infection, alcohol and NSAID ingestion, stress (e.g. severe trauma), or Crohn's disease ► they heal without scarring

  Double-contrast barium studies 1–2mm shallow collections of barium ► short linear or serpentine lines or dots of barium
  - *'Complete' (varioloform) erosions:* a complete radiolucent rim of surrounding oedema is present
  - *'Incomplete' erosions:* the oedema halo is lacking

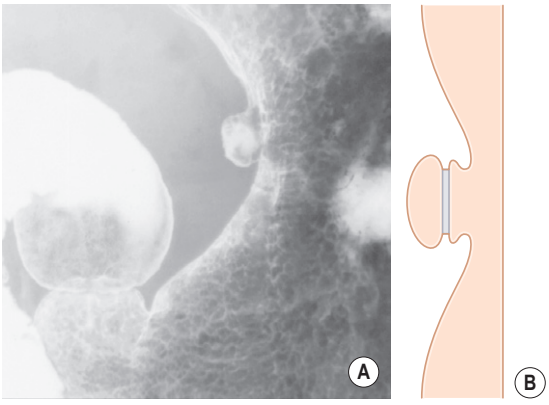


En face appearance of benign gastric ulcer. (A) The posterior wall ulcer is nearly filled with barium in this RPO projection. Thin regular radiating folds (best seen around the inferior border of the ulcer) are seen converging to the ulcer. (B) Unfilled benign ulcer crater is outlined by a 'ring' shadow. This ulcer is surrounded by a prominent ring of oedema – the lucent area around the crater.*

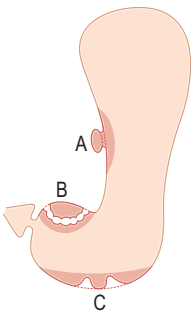




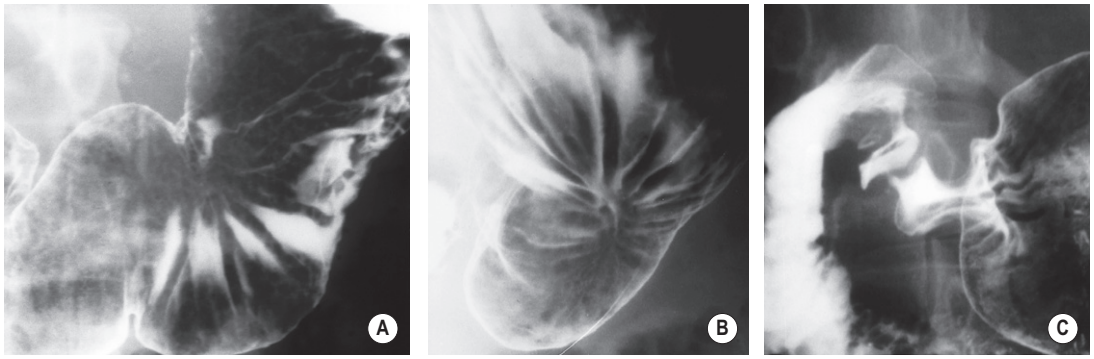
Profile views of benign gastric ulcer. (A) Hampton's line, a thin line of radiolucency crossing the opening of an ulcer, a virtually infallible sign of a benign ulcer. (B) Lesser curvature ulcer with a clearly visible ulcer collar. (C) Large lesser curvature ulcer niche, its projection from the lumen of the stomach strongly suggesting a benign lesion. (D) Smooth, straight radiating folds converge at the ulcer crater.*



Benign gastric ulcer. (A) Mid lesser curvature ulcer in profile. The ulcer crater projects outside the wall of the stomach. (B) Diagram of a benign ulcer with an oedematous collar. Beneath the collar, a thin lucent line (Hampton's line) may be seen across the mouth of the ulcer.†



Graphical description of three types of gastric ulcer (the shading represents barium): A = benign, projecting, lesser curvature ulcer with collar (broken lines = Hampton's line) ► B = malignant, intraluminal ulcer with irregular nodular tumour rim which traps a lenticular barium collection that is convex relative to the gastric lumen (Carman meniscus sign) ► C = non-projecting benign greater curvature ulcer.†



Healing gastric ulcer. (A) Focal retraction along the incisura angularis with small residual outpouching is present. Converging smooth folds no longer fill an ulcer crater. (B) Radiating folds converging to a linear scar. (C) Scarred antrum with constriction at site of previous ulcer causing narrowing and deformity.

GASTRITIS DUE TO *HELICOBACTER PYLORI* INFECTION

DEFINITION

- *H. pylori* is a Gram-negative, flagellated, spiral bacterium ► it is more prevalent in developing countries, tending to affect the lower socioeconomic groups
- The gastric cancer risk is 6 times higher in patients with *H. pylori* infection

Pathophysiology The enzyme urease converts urea into ammonia and bicarbonate – the resultant alkaline 'microenvironment' protects the bacterium from the effects of gastric acid ► acute infection initially injures the parietal cells (decreasing gastric acid production) – once parietal cell function recovers, the resultant abnormally high acid output causes antral gastritis and duodenitis

RADIOLOGICAL FEATURES

Barium meal There can be diffuse or focal changes including: thick or nodular gastric folds ► erosions ► ulcers ► antral narrowing ► inflammatory polyps ► prominent areae gastricae

- The radiological signs are non-specific – similar appearances can be seen with gastritis due to NSAID and alcohol ingestion

HYPERTROPHIC GASTRITIS

DEFINITION

- Glandular hyperplasia and increased acid secretion (inflammation is not prominent)
  - The differential includes Ménétrier's disease and lymphoma

RADIOLOGICAL FEATURES

Barium meal Thickened folds (often >10mm) predominantly located within the fundus and body of the stomach (the acid-producing regions) ► prominent areae gastricae – these can be up to 4–5mm in size and more angular and polygonal than their usual round or oval configuration

- There is a high prevalence of duodenal and gastric ulcers

ATROPHIC GASTRITIS

DEFINITION

- Atrophy of the gastric glands with associated histological inflammatory changes ► it is associated with pernicious anaemia and is more common with advancing age
  - *Pernicious anaemia:* this is caused by decreased intrinsic factor and vitamin B_{12} production (characterized by parietal and chief cell loss leading to achlorhydria and atrophy of the mucosa and mucosal glands)
    - 90% of patients will also have atrophic gastritis
    - There is also an association with gastric polyps, carcinoma, benign and malignant ulcers
- It is also associated with intestinal metaplasia (which is a premalignant condition) – radiographically this is suggested by enlargement of the areae gastricae

RADIOLOGICAL FEATURES

Barium meal A loss of the rugal folds (± the areae gastricae) ► a tubular and featureless narrowed stomach

OTHER TYPES OF INFECTIOUS GASTRITIS

Granulomatous infections

- Including tuberculosis, histoplasmosis and syphilis

Barium meal Ulceration ► thick folds ► mucosal nodularity ► antral narrowing is a late finding

Moniliasis

- This is usually associated with severe oesophageal disease

Barium meal Prominent aphthous ulcers

Immunocompromised patients

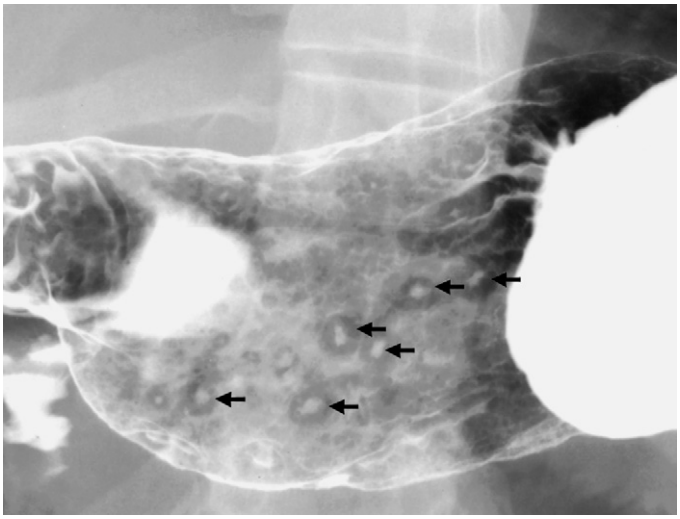
- Infection with cytomegalovirus (CMV), toxoplasmosis, or cryptosporidiosis
  - Cryptosporidiosis primarily affects the small bowel rather than the oesophagus ► it causes severe diarrhoea with thickened small bowel folds

Barium meal Deep ulceration and fistulization (CMV) ► antral narrowing and rigidity (cryptosporidiosis)

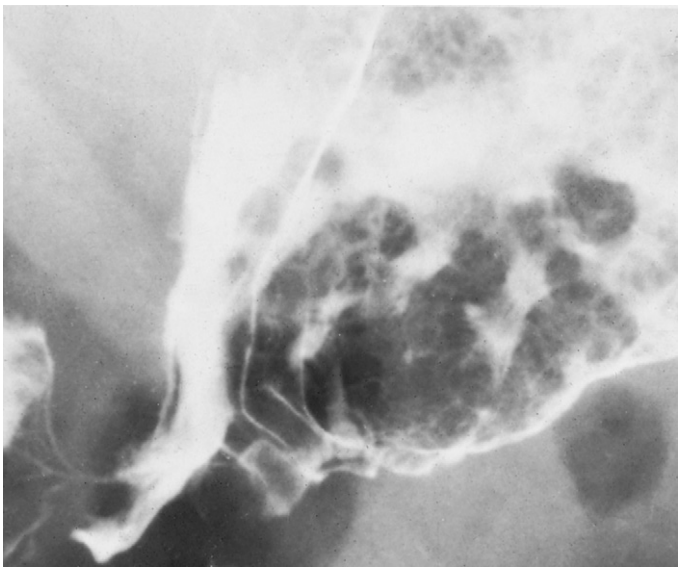
Strongyloides

- A parasitic infection affecting the upper gastrointestinal tract, duodenum and proximal small bowel

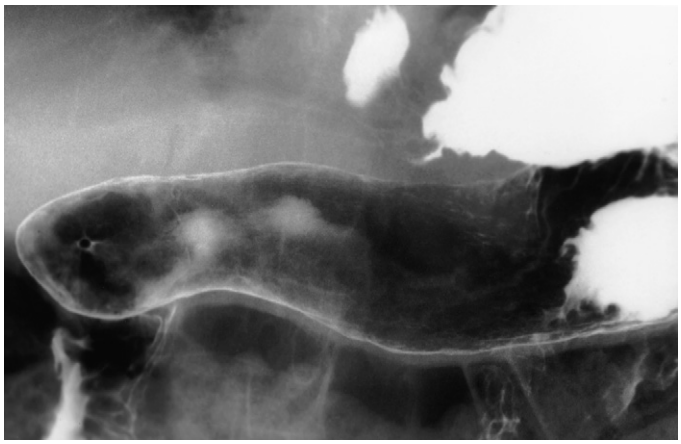
Barium meal Advanced cases will cause thickened effaced folds with associated narrowing



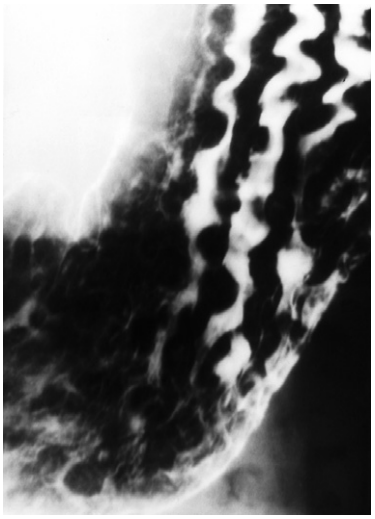
Acute erosive gastritis. There are numerous erosions in the stomach (arrows). Each erosion consists of a small central collection of barium surrounded by a translucent ring (a small 'target' lesion).†



Severe antral gastritis. Conical narrowing of the antrum with multiple thickened gastric folds.†



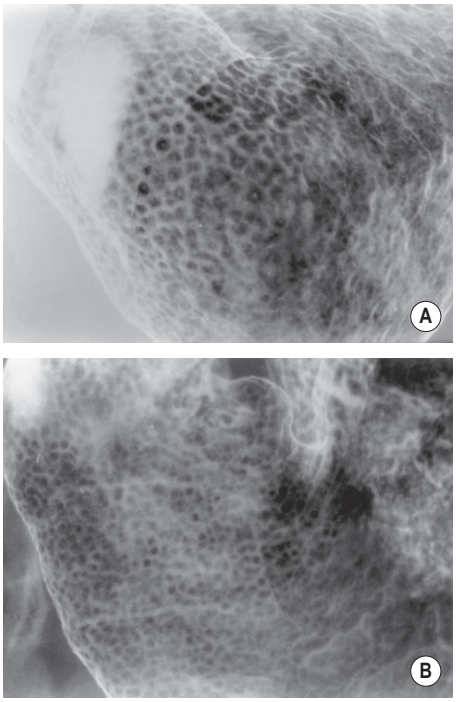
Atrophic gastritis. Featureless narrowed stomach. Note pyloric channel seen 'end on'.*



Diffuse erosive gastritis with thick nodular folds. Erosions are scattered along the folds.*

Prevalence of *Helicobacter pylori* infection with upper GI disease*

Disease	Prevalence (%)
Active chronic gastritis	100
Duodenal ulcer	95
Gastric cancer (body or antrum)	80–95
MALT lymphoma	90
Gastric ulcer	60–80
Non-ulcer dyspepsia	35–60
Asymptomatic population	20-55



H. pylori gastritis with lymphoid hyperplasia. (A) and (B). In both patients, enlarged lymphoid follicles are seen as innumerable tiny, round nodules that carpet the mucosa of the gastric antrum. In (A) note how many of the nodules have central umbilications with punctate collections of barium seen en face in the lesions.©20

GRANULOMATOUS GASTRITIS

DEFINITION

- Granulomatous inflammation of the gastric mucosa can be secondary to: Crohn's disease ► sarcoidosis ► tuberculosis ► syphilis ► fungal disease

PEARL

Crohn's disease

- Gastroduodenal involvement is seen in up to 20% of patients (usually with an associated ileocolitis) ► if the upper GI tract is involved both the stomach and duodenum are usually involved (isolated duodenal involvement is more common than isolated stomach involvement) ► the radiographic findings of gastric disease usually always involve the stomach antrum and body ► a gastrocolic fistula is a rare complication (usually involving the transverse colon)
  - *Early (non-stenotic) phase:* aphthous ulcers ► larger discrete ulcers ► thickened and distorted folds ► a nodular 'cobble-stoned' mucosa
    - These features are indistinguishable from aphthous ulcers or erosions due to other causes
  - *Late (stenotic) phase:* a 'rams-horn' or 'pseudo post-Bilroth I' appearance: this is caused by scarring and fibrosis of the gastric antrum and pylorus – it can foreshorten the stomach enough to simulate a partial gastrectomy
    - This appearance can also be seen with other granulomatous disorders such as tuberculosis, syphilis, sarcoid and eosinophilic gastroenteritits
    - The antral narrowing can also mimic a scirrhous gastric carcinoma

ZOLLINGER–ELLISON SYNDROME

DEFINITION

- A gastrin-secreting gastrinoma (a non-beta islet cell tumour) stimulates excessive gastric acid secretion ► this leads to prominent ulcer formation, often in locations distal to the normal ulcer distribution
  - *Ulcer location:* duodenal bulb > stomach > post-bulbar duodenum
  - *Gastrinoma location:* pancreas (75%) ► duodenum (15%)
  - *Gastrinoma metastases:* these are primarily to the liver (up to 50% of tumours are malignant)
  - 10% of tumours are associated with the type 1 multiple endocrine neoplasia (with associated parathyroid, pituitary and adrenal tumours)

RADIOLOGICAL FEATURES

Barium meal Thickened gastric and duodenal folds ► single or multiple ulcers (10%) ► reflux oesophagitis

EOSINOPHILIC GASTRITIS

DEFINITION

- This follows a focal or diffuse infiltration of the GI tract with eosinophils ► it is associated with atopy, asthma and often a peripheral eosinophilia (there is possibly an atopic aetiology)
  - Any segment of the GI tract can be affected ► however, it commonly involves the stomach (particularly the antrum) and the proximal small bowel

CLINICAL PRESENTATION

- Crampy abdominal pains ► diarrhoea ► distension and vomiting

RADIOLOGICAL FEATURES

- These will depend upon which layers of the GI tract are affected (involvement may be predominantly mucosal, muscular or serosal) ► if the disease is panmural then an eosinophilic ascites is often seen
  - *Oesophagus:* involvement can result in stricture formation
  - *Stomach:* thickened folds ► antral narrowing and rigidity ► mucosal nodularity ► antral and pyloric stenosis is common
  - *Small bowel:* fold thickening ► bowel stenosis

CORROSIVE GASTRITIS

DEFINITION

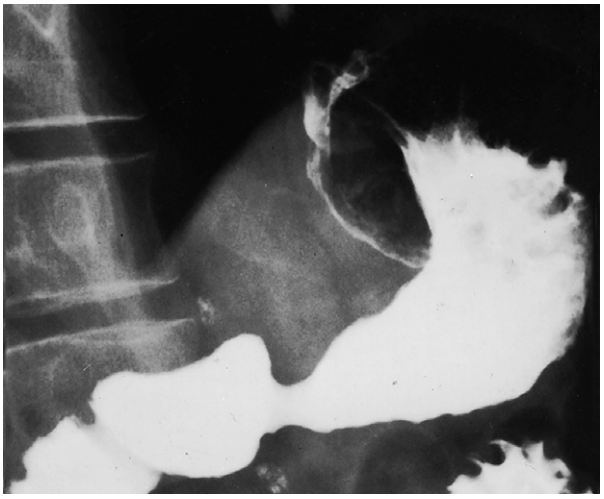
- Inflammation of the gastric mucosa due to acid or alkali ingestion ► acids are more injurious (the already acidic gastric contents have no residual ability to neutralize strong acids)
  - *Initially*: there is necrosis with sloughing of the mucosal and submucosal layers
  - *Moderate cases:* there is subsequent fibrosis and stricture formation – the resultant contracted (± obstructed) stomach may require a gastrectomy
  - *Severe cases*: there can be subsequent full-thickness necrosis with perforation

RADIOLOGICAL FEATURES

Barium meal A swollen, irregular gastric mucosa (occasionally with visible blebs)

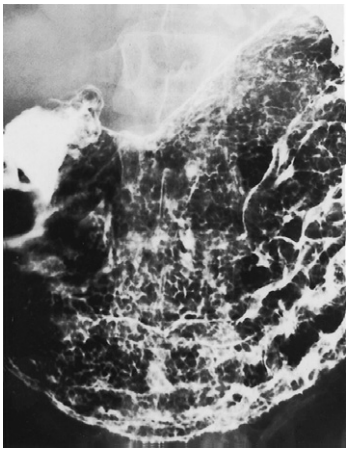
- As this sloughs barium can flow beneath it (the mucosa is then seen as a thin radiolucent line paralleling the outline of the stomach)
- After a few weeks fibrotic contraction of the stomach can occur – this can be severe enough that the stomach lumen is no larger than the duodenal bulb

CT shows diffuse thickening of the gastric wall in a patient proven to have eosinophilic gastroenteritis. No ascites was present. Symptoms resolved with steroid therapy.*

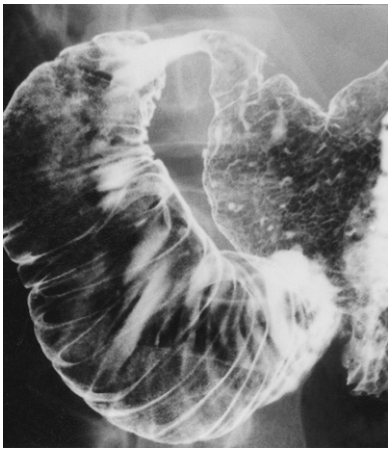


Corrosive gastritis following the ingestion of household bleach. The distal stomach has undergone considerable scarring and contraction in a manner similar to syphilitic gastritis or linitis plastica.*

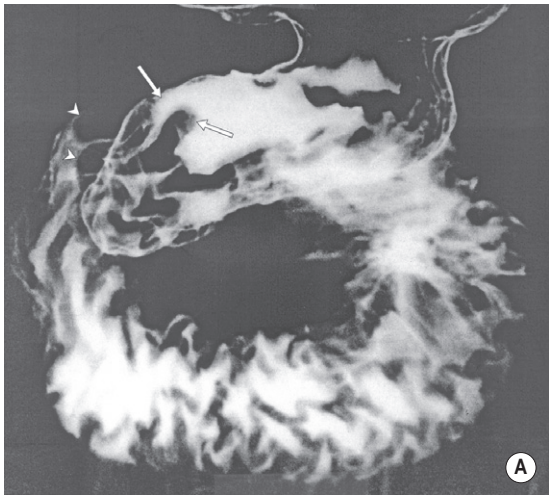
Crohn's disease. Multiple aphthous erosions are present on the antrum. Duodenal folds are thick and nodular.*

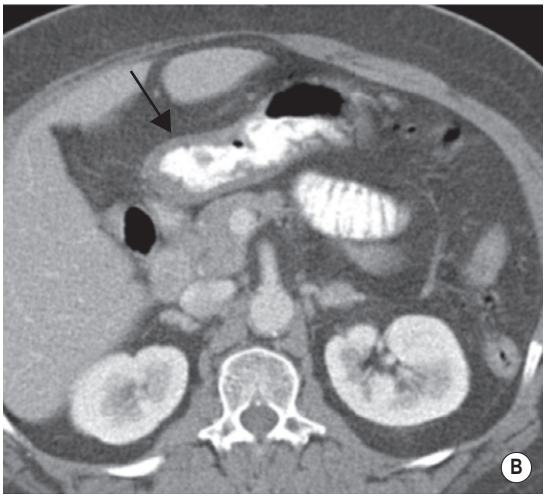


Hypertrophic gastritis in a patient with a recently healed lesser curvature gastric ulcer. This characteristic enlargement and prominence of the areae gastricae can be correlated with an increased incidence of gastric hypersecretion and peptic ulcer disease (PUD).*



Crohn's disease. Antral erosions and a tapered stricture involving the first part of the duodenum. The second part of the duodenum is dilated as a result of a further stricture of the third part.†





(A) Prominent thickened antral folds in a patient with antral gastritis. (B) Axial CT in this patient shows thickened antral wall (arrow) secondary to inflammation.••

BENIGN TUMOURS

DEFINITION

Mucosal polyps

Hyperplastic polyps The commonest benign gastric neoplasm (accounting for 80% of all polyps) ► there is no premalignant potential but they do occur more commonly in patients with other risk factors for gastric malignancy (e.g. atrophic gastritis or bile reflux gastritis)

- *Fundic gland polyp:* a variant representing a hyperplastic fundal gland (therefore they are not found in the antrum) ► they can be seen in up to 40% of patients with familial adenomatous polyposis coli (FAPC)

Adenomas This is a premalignant neoplasm that may develop into a gastric carcinoma (malignancy is detected histologically in 50% of adenomas that are >2cm) ► it is also often found in FAPC (along with hypertrophic polyps)

Hamartomas/inflammatory polyps Along with hyperplastic polyps, these are found in various polyposis syndromes (e.g. Peutz–Jeghers, Cronkhite–Canada, and Cowden's disease)

Submucosal lesions

Gastrointestinal stromal tumour (GIST)

- A benign mesenchymal tumour arising within the submucosa (and previously designated as a leiomyoma, leiomyoblastoma or leiomyosarcoma)
- 70% of GISTs occur within the stomach, accounting for 1–3% of all gastric malignancies ► 70–90% of GISTs are benign
- See Section 3 Chapter 4, Small bowel

RADIOLOGICAL FEATURES

Barium meal

Hyperplastic polyps A round, smooth sessile lesion ► they are usually multiple and of a uniform similar size (5–10mm) ► they are commonly found within the fundus or body of the stomach

- They can rarely present as an isolated large, irregular lesion

Adenoma A polypoid sessile or pedunculated lesion ► they are usually solitary and >1cm in size ► they are commonly found within the antrum

- Villous adenomas can have frond-like projections and are associated with a very high risk of malignancy

MALIGNANT TUMOURS

DEFINITION

Histological types

Gastric carcinoma See Section 3 Chapter 4, Small bowel, Gastrointestinal stromal tumours

Lymphoma

- Lymphoma is usually due to primary disease, as a result of direct extension from adjacent lymph nodes, or as part of a generalized disease process
- The GI tract is the commonest site of primary extranodal lymphoma, with the stomach the most frequent site of a GI lymphoma (accounting for 3% of all gastric malignancies) ► most lymphomas involving the stomach are of the non-Hodgkin's type
  - *MALT lymphoma*: a type of non-Hodgkin's lymphoma occurring within the stomach (its commonest location), lung, thyroid, salivary glands and intestine ► it usually arises within the mucosa-associated lymphoid tissue which has been acquired in response to a *H. pylori* infection (there is normally no lymphoid tissue present within the gastric mucosa)
  - *Hodgkin's type*: this mimics a scirrhous carcinoma (with a strong associated desmoplastic reaction)

GIST Suspect malignancy if it is >5cm in size ► see Section 3 Chapter 4, Small bowel

Metastases

- The most common primary tumours to metastasize to the stomach are breast, malignant melanoma, and lung
- There can also be contiguous spread from the colon (via the gastrocolic and gastrosplenic ligaments), the liver (via the gastrohepatic ligament) or the pancreas (direct spread)

RADIOLOGICAL FEATURES

Lymphoma

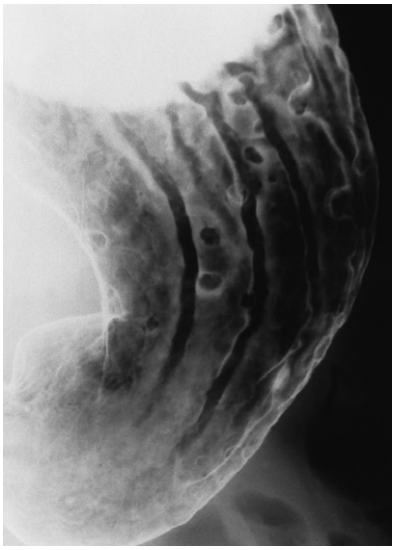
Barium meal/CT There is no typical imaging appearance and it may mimic the appearances of any gastric carcinoma

- The most common appearance is of an infiltrating lesion extending over a large area of the stomach with diffuse gastric fold thickening (± ulceration) ► it may also appear as a bulky polypoid mass or a malignant ulcer
- Lymphoma is more likely to spread across the pylorus and into the duodenum than is a gastric carcinoma
- Direct spread of disease or invasion of the stomach from enlarged regional lymph nodes may be a helpful sign

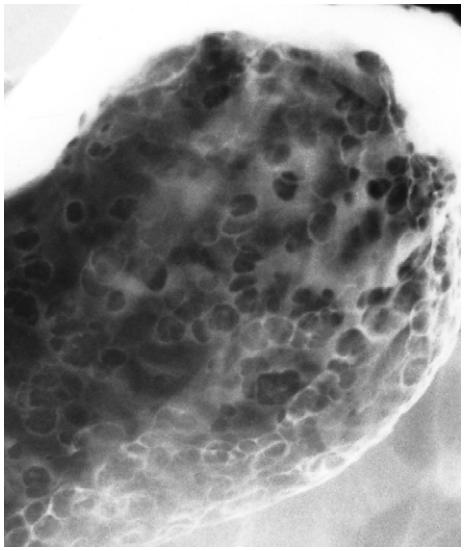
Haematogenous metastases

Barium meal Initially there will be small intramural masses (these may have central ulceration and are most frequently seen with metastatic melanoma, lymphoma, and Kaposi's sarcoma)

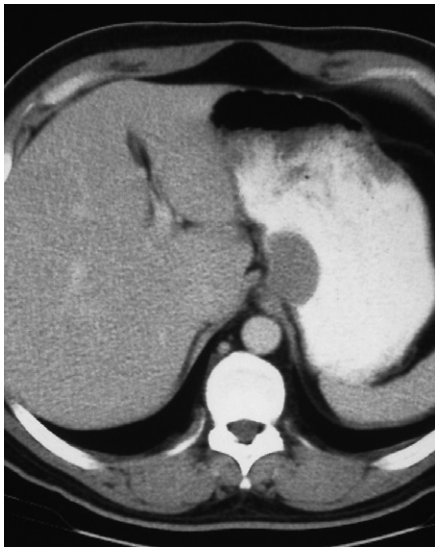
- Breast carcinoma may produce a linitis plastic type appearance (which is indistinguishable from a primary gastric carcinoma)



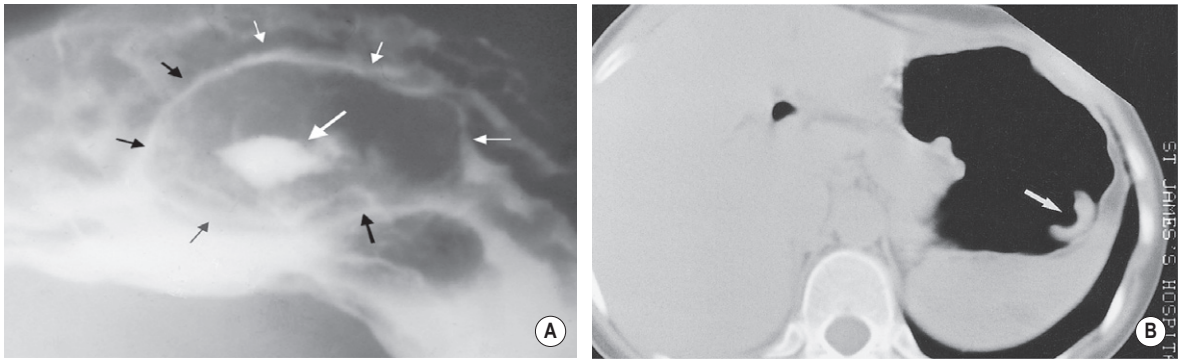
Hyperplastic polyps in the body of the stomach – small, sessile and uniform in size.*



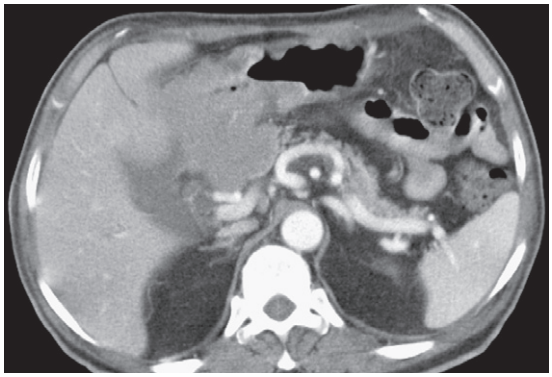
Fundic gland polyps morphologically identical to hyperplastic polyps predominate in the fundus as shown in this image. This is a patient with familial adenomatosis coli.*



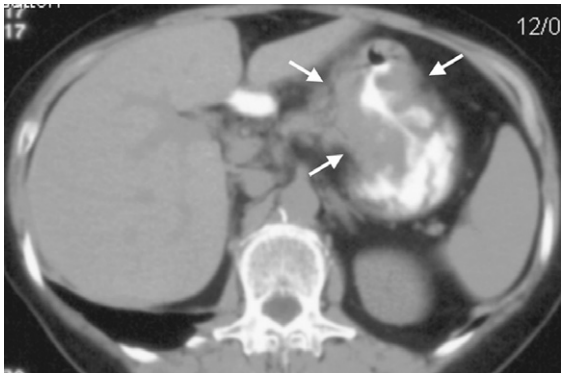
Leiomyoma adjacent to the gastro-oesophageal junction shown on CT as a smooth soft tissue mass in the contrast-filled stomach.*



Gastric 'target' lesion. (A) An ulcerating (large arrow) tumour in the gastric fundus (small arrows). This appearance is typical of an ulcerating submucosal metastasis from a malignant melanoma. (B) CT shows the same tumour (arrow).†



Gastric lymphoma. CECT. Note the marked thickening of the gastric antrum.*



MALT lymphoma. Multifocal tumour (arrows) thickening the gastric wall.†

GASTRIC CARCINOMA

DEFINITION

- A malignant tumour arising from the gastric mucosa (it is an adenocarcinoma in 95% of cases)
  - *Japan:* this has the highest prevalence (which is related to dietary factors)
  - *Western countries:* it is usually detected at an advanced stage (due to its non-specific symptoms)
- The disease results from a progression through chronic inflammation (gastritis), to intestinal metaplasia, and then carcinoma
  - There is a demonstrable link between *H. pylori* colonization and a *distal* gastric carcinoma (*H. pylori*, GOR and bile reflux may play a role in gastric *cardia* tumours)
  - The incidence of gastric cardia tumours has increased – the incidence elsewhere has decreased or remained stable
- **Other risk factors:** diet ► familial factors ► smoking ► chronic atrophic gastritis ► pernicious anaemia ► Ménétrier's disease ► a low socioeconomic status ► obesity ► a Billroth partial gastrectomy ► gastric polyps
  - *Gastric polyps:* adenomas have the highest malignant potential (40%) ► hyperplastic and hamartomatous polyps have a low malignant potential
- **Histological types:**
  - *Diffuse type (40%):* the undifferentiated form ► signet ring cells are present ► there is a worse prognosis and greater dissemination
  - *Intestinal type (60%):* the differentiated form (tubular or papillary glands are present)

RADIOLOGICAL FEATURES

Barium meal

- **Early gastric carcinoma** (confined to the gastric mucosa and submucosa without muscle layer invasion)

Ulcers*		
Findings	**Benign**	**Malignant**
Hampton's line	Present	Absent
Extends beyond the gastric wall	Yes	No
Folds	Smooth, even	Irregular, nodular, may fuse
Associated mass	Absent	Present
Carman meniscus	Absent	Present
Ulcer shape	Round, oval, linear	Irregular
Healing	Heals completely	Rarely heals

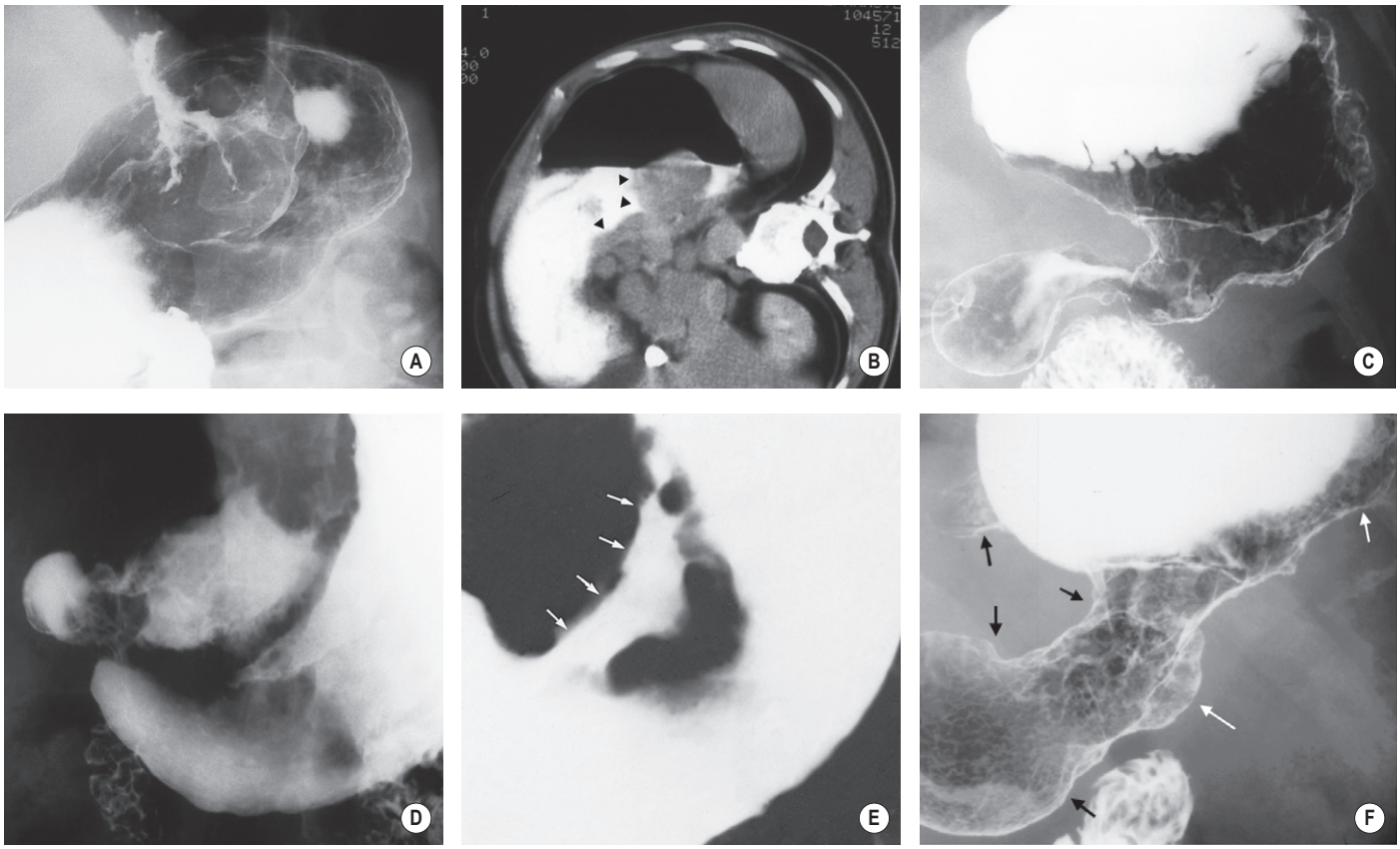
  - A polypoid, superficial or excavated tumour (which is frequently irregular) ► converging folds (which are often thickened, irregular or nodular) ► nodularity is frequently seen around the central portion of a lesion
- **Advanced gastric carcinoma**
  - A large irregular mass (± ulceration) ► an irregular mucosal surface ► the mass margin may exhibit a shelf (forming an acute angle with the gastric wall)
  - Antral involvement can lead to narrowing and obstruction
  - *A 'malignant ulcer':* this indicates an ulcer within a gastric mass
  - *Linitis plastica:* diffuse stomach infiltration with tumour and fibrosis, resulting in a narrowed rigid stomach

CT

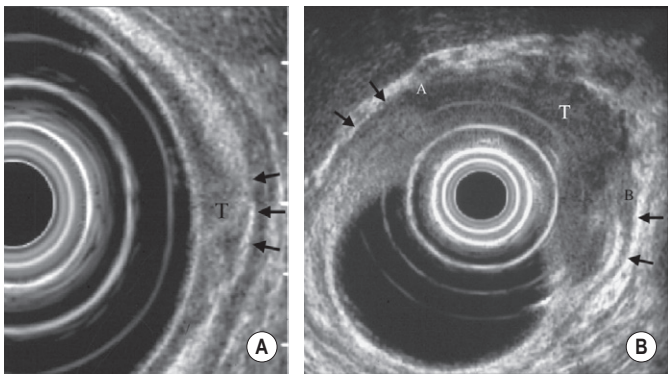
- This requires good distension with water
- **Abnormal signs:** focal wall thickening (± ulceration) ► a focal mass or diffuse wall thickening
  - A wall thickness > 1cm is considered abnormal in a well-distended stomach (except at the GOJ where the transverse imaging plane complicates assessment)
  - There may be abnormal contrast enhancement of the gastric wall or loss of the normal multilayered wall pattern
  - Linitis plastica may complicate assessment due to the associated difficulties with gastric distension
  - Serosal involvement is indicated by an irregular border to the external gastric wall (± perigastric fat stranding)
- **Metastatic spread**
  - *Direct invasion:* involving the pancreas, left liver lobe, spleen, or transverse colon
  - *Haematogenous spread to the liver:* this is seen with 25% of cases at presentation
  - *Intraperitoneal seeding:* to the rectosigmoid colon, caecum, and small bowel ► ascites can also be present
  - *Kruckenberg tumours:* bilateral drop metastases to the ovaries (especially with the signet ring cell tumour type)

EUS

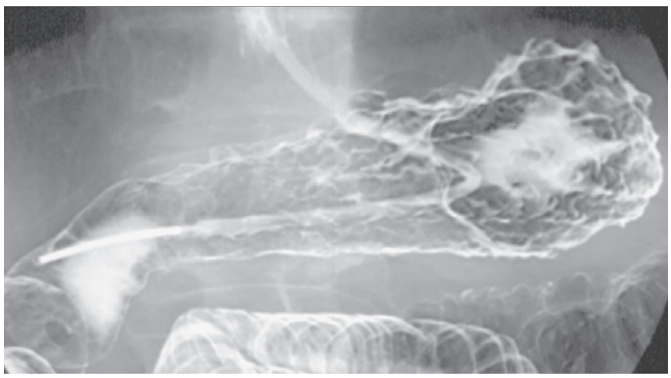
- This is able to resolve the individual layers of the gastric wall, allowing the determination of the T staging (more accurately than with CT)
  - Carcinoma appears as a hypoechoic lesion with irregular margins
- *Nodes:* it can assess the perigastric nodes only (due to its limited depth of view of approximately 6cm) ► it will allow FNA biopsy of any affected nodes



Advanced gastric cancer. (A) Large polypoid mass of the cardia. (B) Polypoid mass of the cardia shown on CT (arrowheads). (C) Large circumferential mass in the body of the stomach with a shelf at the proximal margin sharply demarcating the cancer from the proximal stomach. (D) Large ulcerated mass in the antrum. This is often referred to as a 'Carman' ulcer. (E) Malignant gastric ulcer. Single contrast examination. The ulcer is situated close to the lesser curvature and near the incisura. The arrows indicate the base of the ulcer, which is in line with the lesser curvature, i.e. the crater is non-projecting. Tumour at the margin of the crater appears translucent and nodular, creating a pool of barium, convex one side and concave the other (arrows) ('meniscus' sign). (F) Infiltrating and ulcerative gastric carcinoma. The proximal half of the stomach is involved with thickening of the wall, destruction of mucosa, and narrowing of the lumen (black arrows). Ulceration is present on the greater curve (white arrows).*



Gastric carcinoma. (A) T1 stage. The echogenic submucosal layer has not been breached (black arrows) by the tumour (T). (B) Tumour stage T3. Tumour (T) has breached muscularis propria between points A and B. Intact muscularis propria can be seen at the margins of the tumour (black arrows).†



Small stomach as a result of diffuse submucosal infiltration (linitis plastica).

GASTRIC CARCINOMA

PEARLS

- Early gastric cancer (confined to the mucosa or submucosa) is associated with significant lymph node involvement at presentation (up to 15% of cases) ► the degree of lymph node involvement increases with the depth of submucosal invasion

Lymph nodes Staging is dependent on the number rather than the location of any involved nodes ► a node is considered abnormal if it measures ≥ 8mm along its short-axis diameter (the usual issues regarding the ability to differentiate between enlarged benign reactive nodes and non-enlarged metastatic nodes of course remain):

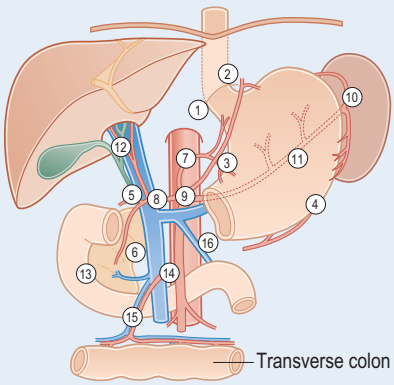
- Compartment III and IV nodes are considered distant metastases (except for the splenic arterial nodes)
- **Surgical dissection:**
  - *D1 lymphadenectomy:* compartment I
  - *D2 lymphadenectomy:* compartments I–II
    - A D2 resection confers improved survival over a D1 resection (but with an increased morbidity)
  - *D3 lymphadenectomy:* compartments I–III
  - *D4 lymphadenectomy:* compartments I–IV

Staging Preoperative tumour staging is usually by CT, and occasionally by EUS for evaluating the depth of any gastric wall invasion ► the role of FDG PET in locoregional staging is limited at present – it is not possible to identify adjacent regional nodes separate to tumour

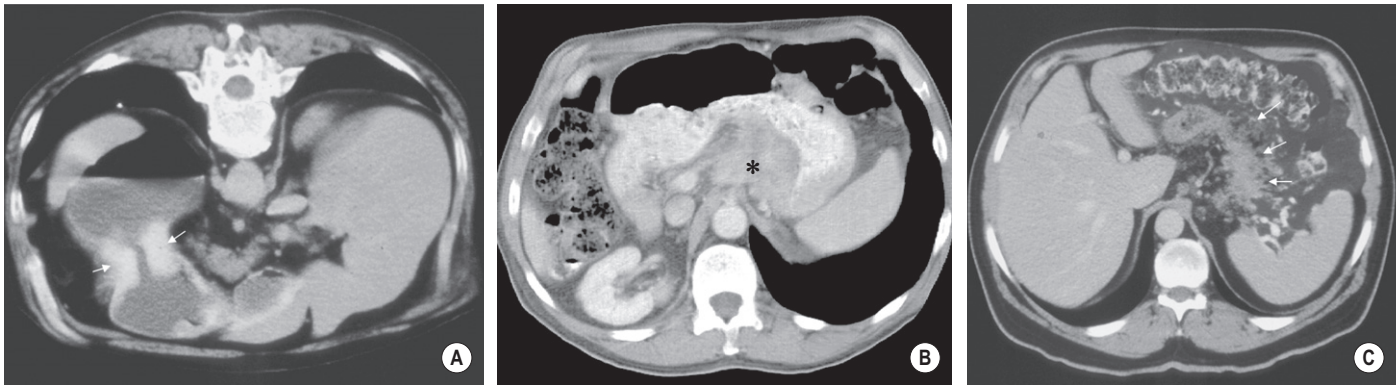
Treatment and prognosis

- Two important factors influence the survival rates for resectable gastric cancer:
  - The depth of invasion
  - Whether there is regional lymph node involvement (compartment II involvement is associated with a worse prognosis)
- **Resectable tumours:** T1, T2 or T3 tumours (without metastases) ► a single resectable liver metastasis may permit possible resection
- **Treatment approach:** gastrectomy (total or subtotal depending on site of tumour) ± lymphadenectomy (limited or extended) ± neoadjuvant chemotherapy
  - Although gastric tumours are chemosensitive there is a minimal impact on long-term survival rates
  - The high incidence of local recurrence, even after apparently complete resection, contributes to the poor long-term prognosis (with a 5-year survival rate of 5%)
  - >50% of patients present with an unresectable locally advanced tumour or metastatic disease ► options then include palliative surgery (± chemotherapy)

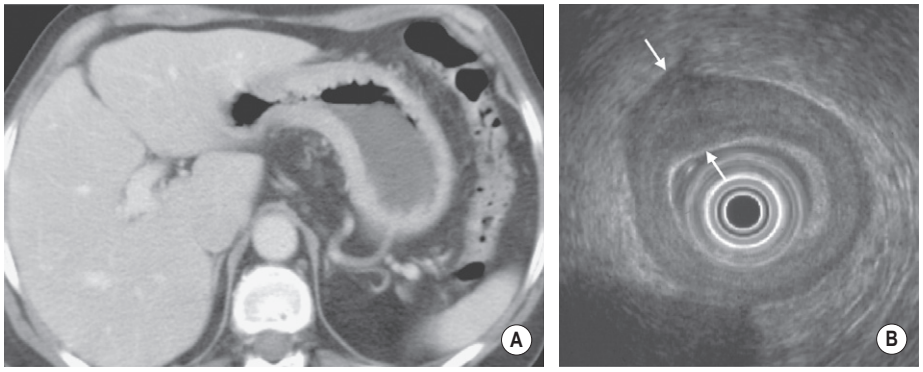
Nodal locations	
Compartment I (perigastric)	Pericardial (left or right) ► lesser or greater curvature ► supra- or infrapyloric
Compartment II	Left gastric ► common hepatic, coeliac or splenic arterial ► splenic hilum
Compartment III	Hepatoduodenal ligament ► posterior to the pancreatic head ► mesenteric root ► splenic artery (if the tumour is within the lower ⅓ of the stomach)
Compartment IV	Middle colic vessels ► para-aortic ► retrocrural



Drawing illustrates lymph node locations according to the Japanese Research Society for Gastric Cancer, *1* = right paracardium, 2 = left paracardium, 3 = lesser curvature, 4 = greater curvature, 5 = supraylorum, 6 = infrapylorum, 7 = left gastric artery, 8 = common hepatic artery, 9 = coeliac artery, 10 = splenic hilum, 11 = proximal splenic artery, 12 = hepatoduodenal ligament, 13 = posterior surface of the pancreatic head, 14 = superior mesenteric vessels (SMA = superior mesenteric artery, SMV = superior mesenteric vein), 15 = middle colic vessels, 16 = abdominal aorta (modified from RadioGraphics 2006 ► 26:143–156).



(A) Gastric carcinoma constricting the stomach body (arrows). Stomach distended with water. Prone image demonstrating that the fat plane between the stomach and pancreas is preserved, excluding pancreatic invasion. (B) Gastric carcinoma (asterisk) extending beyond the serosa to encase the coeliac axis vessels. (C) Extension into the transverse mesocolon (arrows) from an antral carcinoma.†



Linitis plastica – (A) diffuse gastric wall thickening demonstrated on CT. (B) EUS demonstrates diffuse thickening of all layers of the gastric wall (between arrows).†

Gastric carcinoma. The tumour is enhancing and thickening the wall of the antrum (arrows). The stomach is distended with food debris as a result of gastric outlet obstruction.†

Kruckenberg tumours. Bilateral partly cystic ovarian tumours and malignant ascites.†

Lymphoma vs gastric carcinoma		
	Lymphoma	**Gastric carcinoma**
Wall thickening	Very thick	Less thick
Perigastric fat planes	Usually preserved	May be obliterated
Regional adenopathy	Common	Common
Extent of adenopathy	May extend below renal vein Large bulky nodes	Does not extend below renal vein Less bulky nodes
Extent	May involve duodenum	Does not commonly involve duodenum

GASTRIC SURGERY

SURGERY TYPES

Billroth

- A partial gastrectomy (historically performed for peptic ulcer disease) consisting of an antrectomy, vagotomy and creation of either a gastroduodenostomy (Billroth I) or gastrojejunostomy (Billroth II)

Gastric bypass surgery

- The creation of a proximal small pouch from the upper stomach with a surgical bypass of the remaining larger distal stomach remnant (the surgical reconstruction allows drainage of both of the stomach segments)
  - *It commonly uses a 'Roux-en 'Y' reconstruction:* the proximal small bowel is divided, with the distal segment attached to the small stomach pouch ► the proximal small bowel segment (draining the distal stomach pouch via the duodenum) is anastomosed to a segment of mid small bowel

Fundoplication

- The gastric fundus is wrapped around the inferior oesophagus to prevent gastro-oesophageal reflux ► it produces a characteristic deformity of the gastric cardia

Partial gastrectomy and gastroenterostomy

- This is performed with either a cholecysto- or choledochojejunostomy for pancreatic carcinoma

COMPLICATIONS

Acute

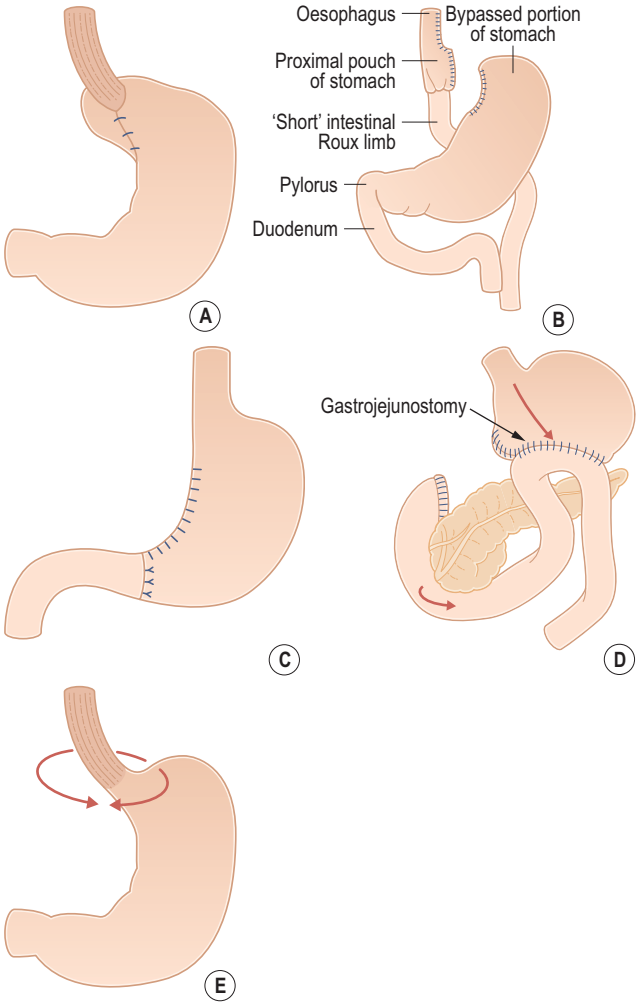
- ***Leakage:*** from the duodenal stump or the anastomosis after gastrojejunostomy is the most common cause of postoperative death
- ***Submucosal haemorrhage:*** this is associated with gastric outlet obstruction (and is self-limiting)
- ***Gastric outlet obstruction:*** this is due to anastomotic oedema
- ***Efferent loop obstruction:*** this is due to spasm or inflammation with delayed transit in the efferent loop (again this is self-limiting) ► it manifests between the 5^{th} and 10^{th} postoperative day

Acute or chronic

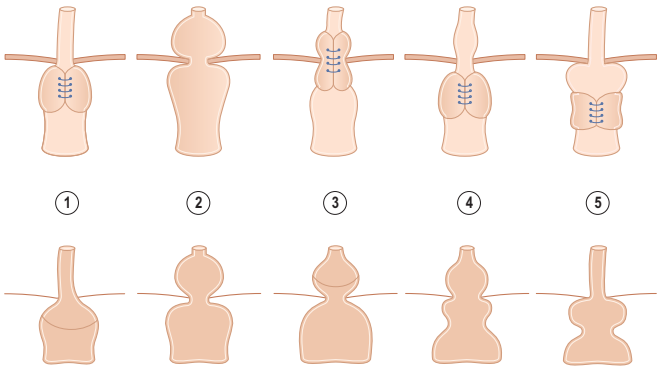
- ***Afferent loop obstruction:*** this follows afferent loop herniation through a surgically created defect behind the gastroenteric anastomosis or because of preferential gastric emptying into the afferent loop ► it presents with intermittent bilious vomiting, weight loss or malabsorption
  - **Barium studies:** there is preferential filling of the afferent loop or afferent loop barium retention on delayed films
- ***Prolapse and intussusception:*** this is commonly jejunogastric, occurring at the anastomotic site (usually affecting the efferent loop)

Chronic

- ***Marginal ulcerations:*** these follow peptic ulcer surgery, and are usually located within 2cm of the anastomosis (on the jejunal side)
  - They are usually caused by an inadequate vagotomy
- ***Phytobezoars (due to a poor diet):*** these can be gastric (following Billroth I surgery) or found within the small bowel (following Bilroth II surgery) ► those within the small bowel may cause obstruction
- ***Primary gastric carcinoma:*** this occurs within the gastric remnant (patients have a relatively high incidence of atrophic gastritis)
  - There is a variable appearance: a lack of distensibility of the gastric remnant ► an intra-luminal mass or ulcer
- ***Gastric outlet obstruction:*** this is due to anastomotic stricture or stenosis formation
- ***Gastric ischaemia leading to necrosis and fistula formation (e.g. gastrogastric or enterocutaneous):*** this is associated with significant mortality
- ***Internal herniation:*** this can be transmesenteric or transmesocolonic
  - The Roux limb or small bowel herniates through a surgical defect within the mesentery or transverse mesocolon

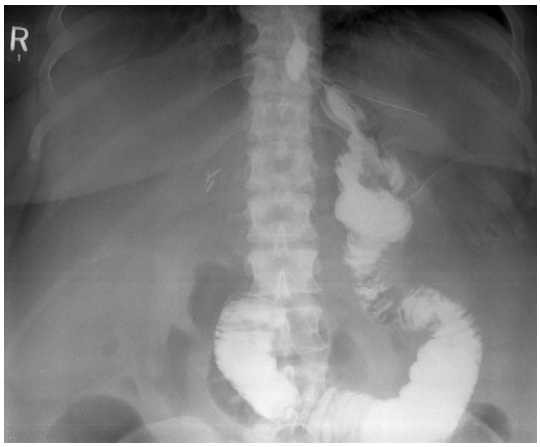


Types of gastric surgery. (A), (E) Nissen fundoplication. (B) Roux-en-Y gastric bypass. (C) Billroth I. (D) Billroth II (gastrojejunostomy).



Appearance of a failure of a fundoplication. Anatomical drawings top row, barium meal appearances bottom row. 1, Normal postoperative appearance ► 2, complete disruption of wrap with recurrence of hiatus hernia ► 3, wrap intact but herniates through diaphragmatic hiatus ► 4, stomach slips up through wrap and bulges above diaphragm ► 5, stomach slips up through wrap but remains below diaphragm.

Bezoar – a large phytobezoar (arrowheads) within the stomach.†



Gastric bypass. Single-contrast upper GI status post gastric bypass.*

Retrograde jejunogastric intussusceptions following gastrojejunostomy. The loops of jejunum within the stomach (arrowheads) have a characteristic 'coiled spring' appearance.†

GASTRIC VOLVULUS

Definition

- The stomach twists on itself between its points of normal anatomical fixation ► it is usually associated with a large sliding or para-oesophageal hiatus hernia and a stomach that is partially or totally within the thoracic cavity
  - **Organo-axial volvulus:** the stomach rotates by 180° along its long axis (a line drawn between the cardia and pylorus)
    - *Complications are rare*
  - **Mesentero-axial volvulus:** the stomach rotates around its short axis (the axis of its mesenteric omental attachments – this is perpendicular to the long axis) ► this is less common but is often associated with a traumatic diaphragmatic rupture
    - *There can be significant clinical consequences*

Clinical presentation

- Violent retching with little vomitus ► severe epigastric pain ► difficulty in passing a NGT
- It may result in gastric outlet obstruction or ischaemia (resulting in a surgical emergency)
- It is most commonly seen in the elderly

Radiological features

Erect XR A double air-fluid level of the stomach within the mediastinum or upper abdomen

Barium meal

- **Organo-axial volvulus:** the greater curvature lies above the lesser curvature (occurring when the original position of the stomach is horizontal) or it is seen as a right-left twist (occurring when the original position of the stomach is vertical)
- **Mesentero-axial volvulus:** an 'upside down stomach' – the distal antrum and pylorus is cranial to the fundus and proximal stomach with the torsed area as a site of obstruction

HYPERTROPHIC PYLORIC STENOSIS

Definition

- Hypertrophy and hyperplasia of mainly the circular muscle results in lengthening and narrowing of the pyloric channel

Clinical presentation

- It is a relatively frequent congenital disorder diagnosed in infancy (commonly affecting first-born males and peaking between 3 and 6 weeks after birth)
- It presents with non-bilious projectile vomiting and a hypokalaemic hypochloraemic metabolic alkalosis

Radiological features

US This usually gives the definitive diagnosis

- Single wall pyloric thickness: > 3mm
- Pyloric length: >16mm
- Transverse pyloric diameter: > 11mm

Barium meal Delayed gastric emptying ► GOR

- *'Tit' sign:* the canal indents the distal antrum
- *'String' sign:* pyloric canal elongation
- *'Shoulder' sign:* the hypertrophied muscle bulges retrogradely into the antrum

Pearl

- **Acquired hypertrophy of the distal antrum and pylorus:** this occurs in peptic or other inflammatory disease in adults ► there is no retrograde muscular bulge

MÉNÉTRIER'S DISEASE

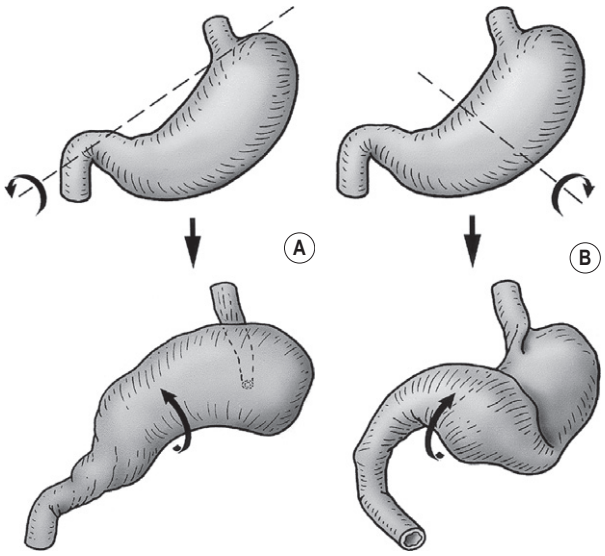
Definition

- This is characterized by gastric gland hypertrophy, achlorhydria or hypoproteinaemia ► it is associated with gastric carcinoma
  - A protein losing enteropathy occurs due to protein loss from the hyperplastic mucosa into the gastric lumen (with an associated increase in small bowel fluid)

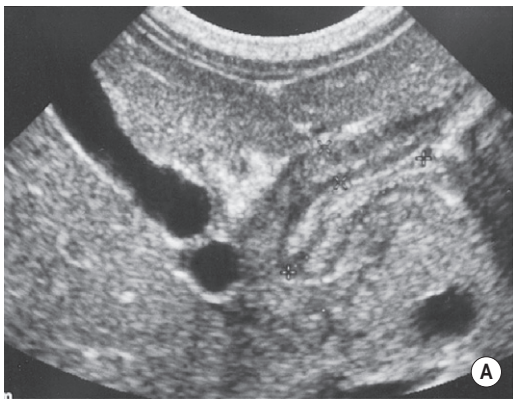
Radiological features

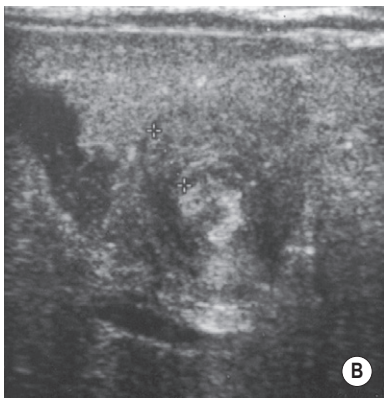
Barium meal Bizarre, markedly enlarged gastric folds (most prominent within the proximal stomach and along the greater curvature) ► poor mucosal coating with barium (due to the increased fluid) ► gastric wall and small bowel fold thickening (due to the hypoproteinaemia)

- Although classically not involved, the antrum can be involved in up to 50% of cases
- The thickened folds remain pliable (cf. a rigid stomach with carcinoma)



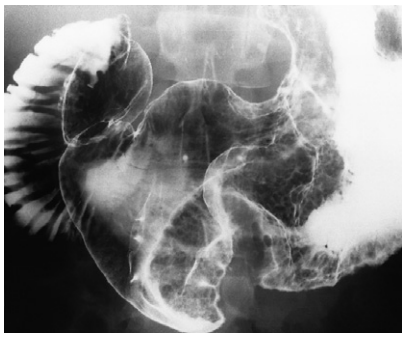
Organoaxial volvulus. (A) Rotation occurs around an axis connecting the pyloris to the oesophagogastric junction, with the greater curve folded upward and to the right. (B) Mesenteroaxial volvulus. Rotation occurs around an axis connecting the middle of the greater curve to the middle of the lesser curve. Generally this type of volvulus is partial as a result of excess mobility of the antrum and duodenum and so the stomach often kinks and obstructs between the body and the antrum.





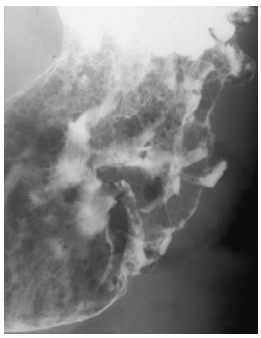
Hypertrophic pyloric stenosis. Longitudinal US image (A) showing an elongated thickened pylorus, muscle length 17mm and width 4mm. Transverse image (B) with muscle width 6mm.†

Hypertrophic pyloric stenosis. Barium meal showing the narrow pyloric canal with a double track of barium. The hypertrophied pylorus indents the base of the duodenal cap.*



Ménétrier's disease. Classic appearance with massively distended folds in the body without abnormality in the antrum.*

Ménétrier's disease. Gross thickening of the folds of the upper two-thirds of the stomach. These patients often weep a protein-rich exudate from the stomach wall, and this excess of fluid in the stomach may impair barium coating.†



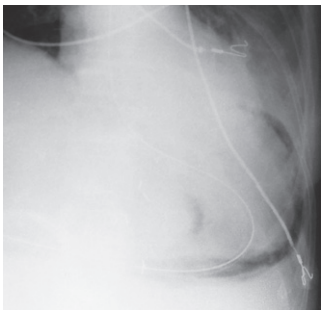
AIR WITHIN THE GASTRIC WALL

Definition

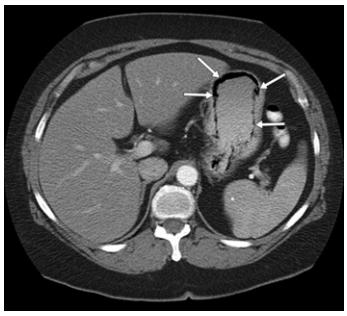
- Disrupted gastric mucosa allows air entry into the gastric wall
  - **'Gastric emphysema':** this occurs without any underlying infection (e.g. secondary to corrosive ingestion, a gastric ulcer, gastric outlet obstruction, COPD, ischaemia, or trauma)
  - **'Emphysematous gastritis':** this is due to an acute infection with a gas forming organism (e.g. *Escherichia coli* or *Clostridium perfringens*)
  - **'Phlegmonous gastritis':** acute panmural infection with a non-gas-forming organism

Radiological features

AXR/CT It is characterized by thin curvilinear lines of radiolucent gas paralleling the gastric wall



Gastric emphysema on AXR in a patient with ischaemic gastritis after extensive abdominal surgery.*



Computed tomographic scan shows air in gastric wall (arrows) after instrumentation.••

PRE-PYLORIC WEB (ANTRAL MUCOSAL DIAPHRAGM)

Definition

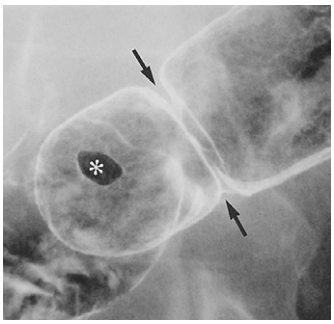
- A congenital thin diaphragm-like, exaggerated fold of gastric mucosa perpendicular to the long axis of the stomach (demarcating the distal antrum into a third small chamber between the proximal antrum and duodenal bulb)

Clinical presentation

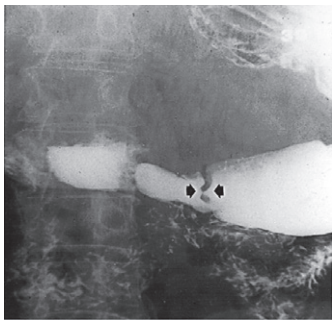
- It can be asymptomatic or can cause gastric outlet obstruction

Radiological features

Barium meal A thin persistent circumferential smooth band within 3-4cm of the pylorus



An antral diaphragm (between arrows). The pyloric canal is seen end on (asterisk).†



Upper gastrointestinal (UGI) study shows a thin, incomplete, web-like structure in the antrum of the stomach representing an antral web (arrows).••

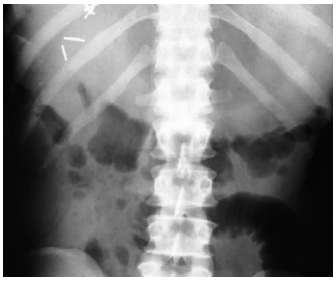
GASTRIC DISTENSION

Obstructive causes

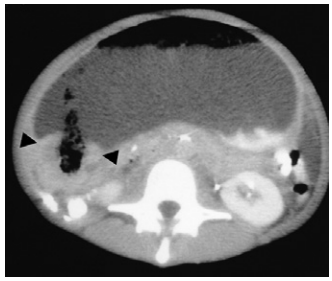
- This is usually due to peptic ulcer disease (duodenal, pyloric, or antral ulcers) ► carcinoma of the antrum or pylorus is the 2nd commonest cause
  - *Rarer causes:* Crohn's disease ► pancreatitis ► pancreatic cancer

Non-obstructive causes

- Paralytic ileus (commonly seen in elderly patients) ► abdominal surgery ► acute trauma ► peritonitis ► chronic diabetes



Supine AXR shows the transverse colon depressed by a distended, fluid-filled stomach. (Surgical clips are present in the right upper quadrant from prior cholecystectomy.)*



Gastric outlet obstruction – cancer of the antrum. Markedly distended stomach with air-fluid level on CT. In this case, a mass in the distal antrum is seen (arrowheads).*

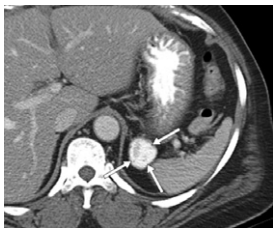
GASTRIC DIVERTICULA

Definition

- This is a true diverticulum (containing muscularis propria) – thus it will demonstrate peristalsis ► it is usually several cm in size and readily fills with barium
  - It is most commonly seen within the posterior aspect of the fundus (near the lesser curvature) ► it can be confused with a left adrenal lesion

Pearl

Intramural or partial gastric diverticula These are due to invagination of the gastric mucosa into the gastric wall ► they are usually <1cm in size with a lenticular shape (in profile) and a small opening into the gastric lumen ► they usually occur along the greater curvature ► they can be mistaken for an ulcer or pancreatic rest ► they are usually asymptomatic



Computed tomographic scan shows a rounded collection of contrast material (arrows) arising from the posterior gastric fundus - this represents a fundal diverticulum.••

On UGI examination, a small, shallow protrusion along the greater curvature of the stomach (arrow) was diagnosed to be a shallow gastric ulcer. Note the absence of inflammatory changes around the area. Endoscopy showed no evidence of inflammation or ulceration and a small antral diverticulum.••

GASTRIC VARICES

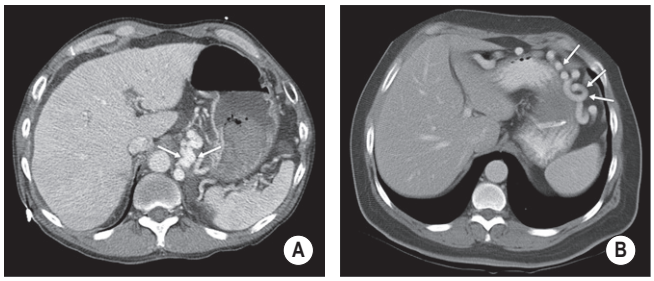
Definition

- These usually occur in patients with portal hypertension and oesophageal varices (as the gastric veins are an additional collateral pathway)
  - The presence of gastric varices, in the absence of oesophageal varices, is a sign of a splenic thrombosis (which is usually associated with pancreatitis or pancreatic cancer)

Radiological features

Barium study/CT Widened effaceable polypoid enhancing folds – these can be nodular-appearing, 'grape-like', or mass-like (the latter mimicking gastric cancer) ► they are usually seen at the fundus and around the GOJ (the antrum is rarely involved without fundal involvement)

- *Differential diagnosis of thick polypoid gastric folds:* hypertrophic gastritis ► Ménétrier's disease ► lymphoma



(A) Large serpentine varices (arrows) are seen in the gastric fundus. (B) Coiled serpentine varices are also seen along this patient's greater curvature (arrows) of stomach.••

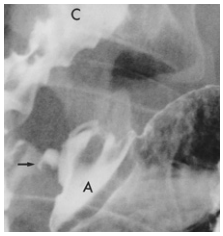
ECTOPIC PANCREAS (PANCREATIC REST/ABERRANT PANCREAS)

Definition

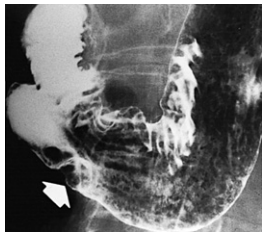
- Pancreatic tissue located within the submucosa of the luminal GI tract
  - This is most commonly seen along the greater curvature of the antrum (also within the 1st and 2nd parts of the duodenum) ► it is usually a solitary lesion

Radiological features

Barium meal A sharply defined submucosal nodule (< 2cm) ► 50% have a central depression or umbilication (representing a rudimentary duct)



Ectopic pancreatic rest. These are generally found in the distal antrum on the greater curve. The small diverticulum results from barium entering the primitive duct system (arrow). A = distal antrum ► C = duodenal cap.†



Marginal polypoid lesion is seen along the greater curvature of the antrum (arrow). Endoscopic evaluation and biopsy specimen showed this to be ectopic pancreatic tissue.••

3.3 DUODENUM

PEPTIC ULCERATION

Definition

- Mucosal ulceration occurring within an acidic part of the GI tract ► it is often associated with *H. pylori* infection ► duodenal ulcers are 2–3x more common than gastric ulcers
  - **Bulbar ulcers** (95%): these are usually benign
    - *Location:* anterior wall > posterior wall
  - **Postbulbar ulcers** (5%): these are usually malignant (95%) and fail to heal on medical treatment
    - *Location:* these are usually seen on the concave border of the 2nd part of the duodenum or within the immediate postbulbar area
- *Risk factors:* surgery ► severe head injury ► steroids ► COPD

Radiological features

Barium studies A duodenal ulcer appears as a sharply defined constant collection of barium (± surrounding oedema or radiating folds)

- *Postbulbar ulcer:* a typical crater is seen – often with spasm of the opposite wall (± thickened mucosal folds and a narrowed lumen) ► scar formation may obscure the ulcer crater
- *Kissing ulcers:* this describes two ulcers opposite each other on the anterior and posterior walls
- *Giant duodenal ulcer:* a benign ulcer crater measuring >2cm ► it is constant in size and shape and often has a sharp round or oval outline ► its floor may be irregular (particularly when the ulcer is penetrating an adjacent organ) ► due to its size it may simulate a deformed duodenal bulb or diverticulum
- *A 'cloverleaf' or 'hourglass' deformity:* this can occur when an ulcer heals with scarring

Pearls

- Multiple postbulbar ulcers occur in the Zollinger–Ellison syndrome
- **Ulcer complications:** perforation ► bleeding ► stenosis ► penetration of any adjacent organs
  - *Perforation:* this can be localized or 'walled off' with marked duodenal deformity due to the adjacent inflammatory reaction

GASTRIC HETEROTOPIA

Definition

- Gastric mucosa occurring in various ectopic locations within the bowel (e.g. the duodenum, small bowel or rectum) ► it is found in a small percentage of normal people

Radiological features

Barium meal Irregular filling defects (varying in size from 1 to 6mm) seen within the duodenal cap, extending from the pylorus distally

Pearls

- This should be differentiated from lymphoid hyperplasia of the duodenal bulb

DUODENAL DIVERTICULA

Definition

- Serosal and mucosal herniations through the muscular wall of the duodenum (seen in 2–5% of barium studies)

Clinical presentation

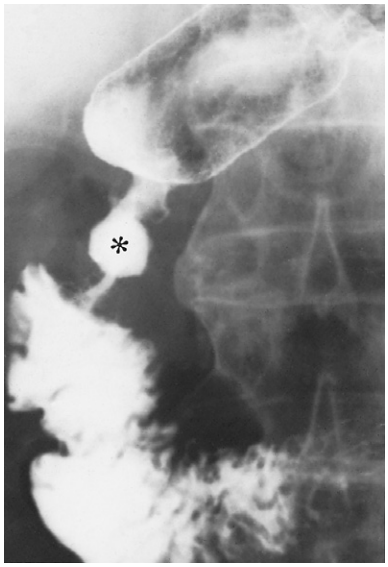
- It is usually an asymptomatic incidental finding (symptoms may occur due to the retention of food or a foreign body) ► it is a rare cause of haemorrhage or perforation
- Occasionally it may contain aberrant pancreatic, gastric, or other functioning tissue – it then becomes a possible site of ulceration, perforation or gangrene
- Cholangitis or pancreatitis may result from the aberrant insertion of the common bile duct or pancreatic duct into an intraluminal diverticulum (with associated impaired biliary drainage)

Radiological features

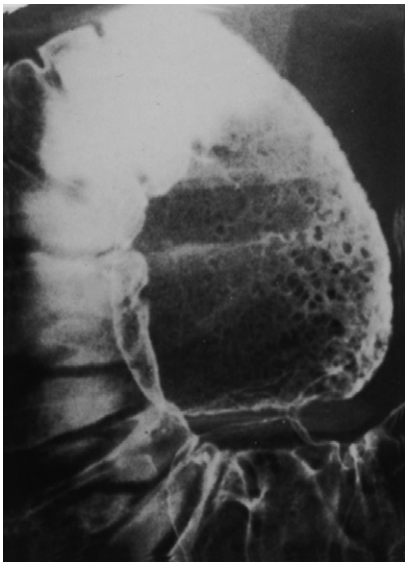
- It is usually found within the 2nd part of the duodenum with most (85%) arising from the medial periampullary surface ► the diverticulum is frequently in contact with the pancreas (and may be embedded in its surface)

Pearl

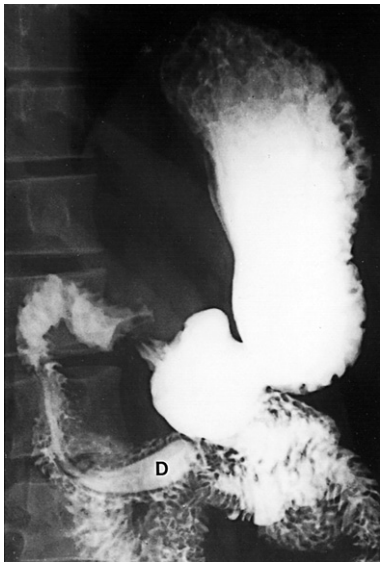
- Duodenal ulceration with spasm or scarring may deform the duodenum, producing a pseudodiverticulum – these are deformable (unlike an ulcer)



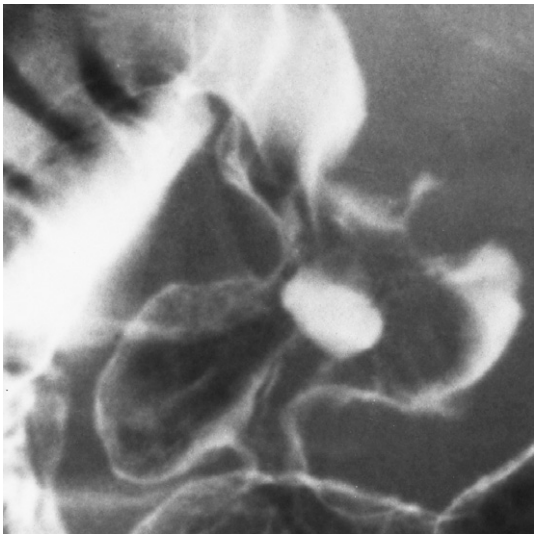
Postbulbar duodenal ulcer. Characteristic appearance with an ulcer crater (asterisk) in the middle of a stricture produced by spasm and oedema.†



Gastric heterotopia. Multiple small irregular filling defects of varying size are seen in the duodenal cap.*

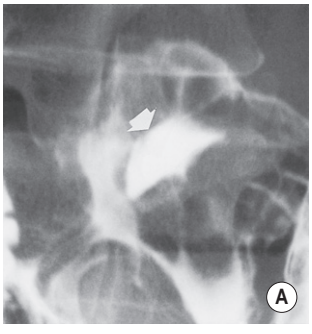


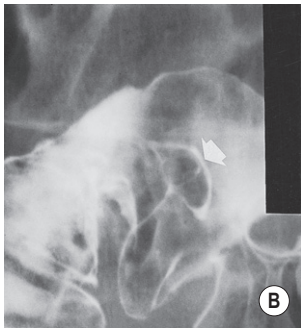
UGI study demonstrates intraluminal diverticulum or wind sock configuration (D).••



Duodenal ulceration. The duodenal cap is deformed and a moderate-sized ulcer crater is outlined with barium.†

Giant duodenal ulcer replacing the duodenal cap.*





Anterior wall duodenal ulcer. (A) prone projection. The ulcer (arrow) is dependent, and so fills with barium. (B) supine projection. The ulcer, which is now on the non-dependent wall of the cap, is outlined with a ring of barium (arrow).†

Duodenal ulcer. Barium collects in an ulcer on the dependent (posterior) wall of the duodenal cap.†

BENIGN TUMOURS

TYPES AND APPEARANCES ON BARIUM STUDIES

Lipoma An intraluminal filling defect (3-4cm in size) which is sharply marginated, solitary, and sessile ► it is easily deformed by peristalsis or compression on fluoroscopy

Brunner's gland hyperplasia These are single or multiple polypoid lesions within the 1st part of the duodenum (often with a characteristic cobblestone appearance) ► a single Brunner's gland adenoma is occasionally seen as a 1cm smooth polypoid mass

- The Brunner's glands normally produce alkaline secretions to protect the duodenal mucosa from gastric acid

Adenomatous polyps Intraluminal filling defects (< 1cm) which can be solitary, sessile or polypoid ► they are seen as a soft tissue mass on CT

Villous adenomas These have a characteristic 'cauliflower' or 'soap bubble' appearance (caused by trapping of barium in the crevices between the multiple frond-like tumour projections) ► they are often 2–3cm in size

Benign lymphoid hyperplasia This is seen as multiple small rounded filling defects of uniform size

- This may be a normal finding in children ► in adults it can be associated with hypogammaglobulinaemia and giardiasis

Carcinoid This is seen especially within the peripapillary region of the 2nd part of the duodenum as a discrete smooth polyp or an irregular infiltrating lesion

Other benign tumours Periampullary adenoma ► gastrointestinal stromal tumour (GIST) ► neurogenic tumour (e.g. neurofibroma) ► hamartoma

PEARL

- Benign duodenal tumours are more common than malignant tumours ► they are often asymptomatic
  - The commonest tumour types are lipomas and leiomyomas

MALIGNANT TUMOURS

TYPES AND APPEARANCES ON BARIUM STUDIES

Primary tumours

Adenocarcinoma of the papilla of Vater This appears as an enlarged papilla of Vater with irregular borders (sometimes with spiculation and ulceration)

- This is the most commonly encountered malignant duodenal tumour ► it usually presents with jaundice

Non-papillary adenocarcinomas of the duodenum These appear as ulcerative, polypoid or annular lesions

- **CT:** focal masses with asymmetric mural thickening and luminal narrowing ► co-incident adenopathy or hepatic metastases may be present
- It usually presents with obstruction

GIST/lymphoma These can also be found within the duodenum

Secondary tumours

Carcinoma or lymphoma of the stomach These can spread directly across the pylorus to involve the duodenum (seen in 25% and 40% of cases, respectively)

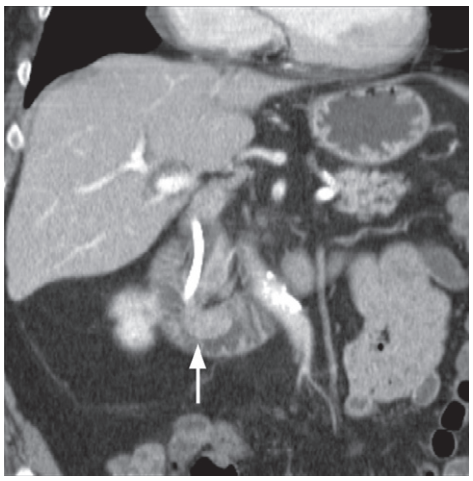
Carcinoma of the head of the pancreas A widened duodenal loop ► a double duodenal contour (± irregularity of the inner border) ► stricturing or distortion of the valvulae conniventes ► a reversed '3' sign of Frostberg

- Carcinoma of the tail of the pancreas may compress or invade the duodenum (resulting in mucosal destruction, bleeding or obstruction)

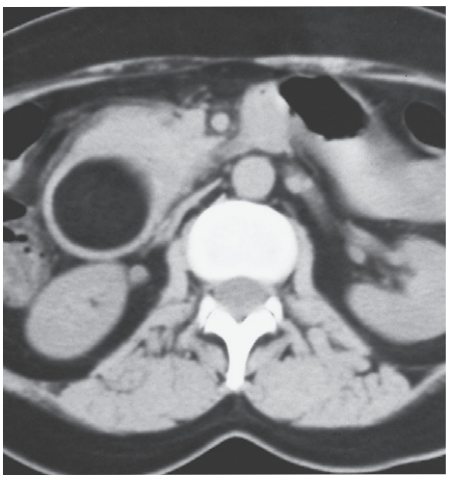
Carcinoma of the colon (particularly the hepatic flexure) This may cause destruction of the mucosal pattern, stricturing, or the formation of a postbulbar ulcer ► a duodenocolic fistula may also form

Carcinoma of the gallbladder This may displace, compress or infiltrate the distal ½ of the 1st part of the duodenum

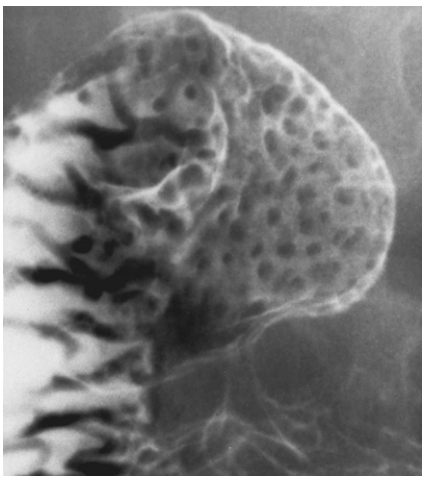
Enlarged neoplastic retroperitoneal lymph nodes These may also invade the duodenum



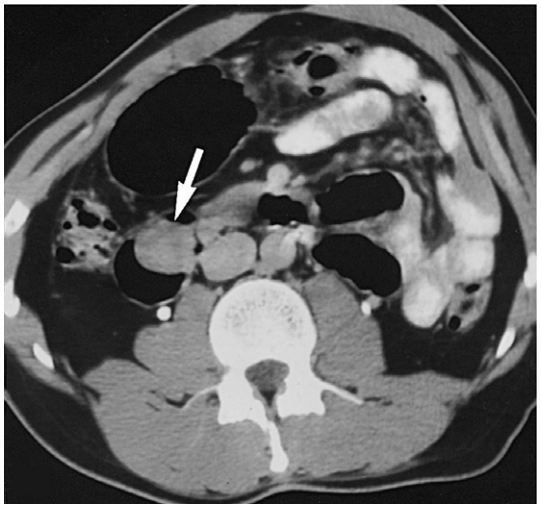
Periampullary adenoma (coronal reconstruction). CECT with water used as oral contrast shows a sharply defined mass (arrow) within the lumen of the second duodenal part extending to the infrapapillary area. A biliary stent is also seen.*



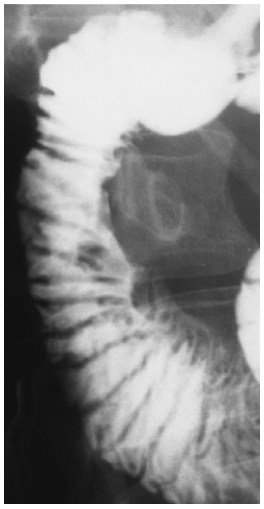
Lipoma. CT shows the lesion to be a well-defined, round mass with low attenuation values, characteristic of fat.*



Lymphoid hyperplasia. Multiple small filling defects characteristic of lymphoid hyperplasia are shown on a double-contrast view of the duodenal cap.*



Ampullary carcinoma. CT scan showing a soft tissue intraluminal mass (arrow) arising from the medial aspect of the descending duodenum.©20



Carcinoma of the pancreas. Barium examination of the duodenum shows the characteristic reversed '3' sign of Frostberg with effacement and distortion of the mucosal pattern on the medial wall of the second portion of the duodenum.*

Primary duodenal adenocarcinoma. CECT with water used as oral contrast shows asymmetric mural thickening and encroachment of the lumen at the second portion of the duodenum (arrow). Dilatation of the common bile duct is also seen. The head of pancreas (P) appears normal.*

CROHN'S DISEASE

Definition Crohn's disease affects the duodenum in 4% of cases ► the radiological appearances are similar to those seen within the more distal small intestine

Barium meal The valvulae conniventes are frequently thickened ► with advanced disease there may be stricture formation with eccentric or concentric narrowing ► cobblestoning, asymmetry and skip lesions may be seen (fissure ulcers, sinuses and fistulae are uncommon)

- *A 'pseudo post-Billroth I' appearance:* this can result from continuous tubular narrowing of the antrum and proximal duodenum

TUBERCULOSIS

Definition This rarely affects the duodenum (if it does it is usually the 2nd part)

Barium meal A narrowed lumen (± mucosal destruction and ulceration)

- Tuberculous mesenteric lymphadenitis (in the absence of intrinsic duodenal tuberculosis) may produce extrinsic pressure on the duodenum leading to obstruction

RADIATION DAMAGE

Definition Duodenal damage is very uncommon – if it does occur, it will usually affect the 2nd part

Barium meal Ulceration ► thickened mucosal folds ► stricturing

PROGRESSIVE SYSTEMIC SCLEROSIS

Definition The duodenum is frequently involved

Barium meal Gross dilatation (which is often more pronounced in the 2nd, 3rd and 4th parts of the duodenum) ► the dilated duodenum can be slow to empty (the atonic organ can produce a sump effect)

INTRAMURAL HAEMATOMA

Definition This is usually due to blunt abdominal trauma

Barium meal Intramural haematoma is usually seen as a concentric obstructive lesion within the duodenum ► thickened valvulae conniventes can result from infiltration by blood and oedema

CT This will demonstrate the extent of any haematoma and will be seen as a mixed attenuation mass surrounding the duodenum

TRAUMATIC RUPTURE

Definition The most frequent site of rupture is at the junction of the 2nd and 3rd parts of the duodenum

CT Retroperitoneal air adjacent to the duodenum ► extravasation of oral contrast medium into the retroperitoneum ► duodenal wall oedema ► peripancreatic fat stranding

SUPERIOR MESENTERIC ARTERY COMPRESSION SYNDROME

Definition This is a rare form of high intestinal obstruction due to narrowing of the normal angle between the aorta and the superior mesenteric artery (through which the 3rd part of the duodenum passes) ► there is a possible association with marked weight loss

Barium meal Strong to-and-fro peristalsis and proximal duodenal dilatation due to compression of the 3rd part of the duodenum ► there will be a sharp cut-off seen in the right anterior oblique position with compression and proximal dilatation persisting when the patient is prone

CT This can measure the angle between the aorta and superior mesenteric artery (on sagittal views)

AORTIC ANEURYSMAL COMPRESSION

Definition An aortic aneurysm may compress the 3rd part of the duodenum and can occasionally cause duodenal obstruction

Pearl A faintly opacified duodenum can stretch around an aneurysm and mimic a contained leak or perianeurysmal inflammation

AORTODUODENAL FISTULA

Definition This is an abnormal communication between the aorta and the duodenum which is usually seen in patients who have undergone aortic graft surgery ► it usually involves the 3rd part of the duodenum

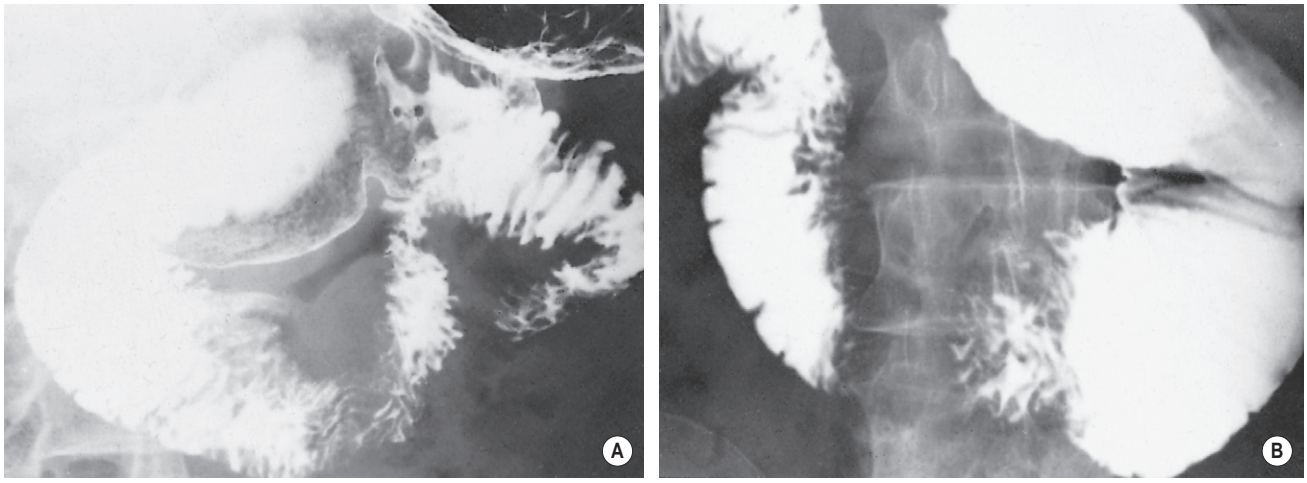
Clinical presentation Patients can present with gastrointestinal haemorrhage

CT Abnormal passage of IV contrast medium from the aorta into the duodenum (endoscopy can also be used for detection)

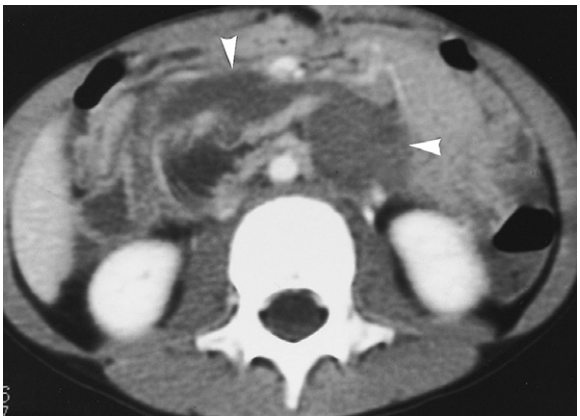
BOUVERET'S SYNDROME

Definition Following erosion of a gallstone through the gallbladder wall into the duodenum, the gallstone may travel retrogradely and become impacted within the duodenal cap (cf. a gallstone ileus with impaction at the terminal ileum following antegrade movement)

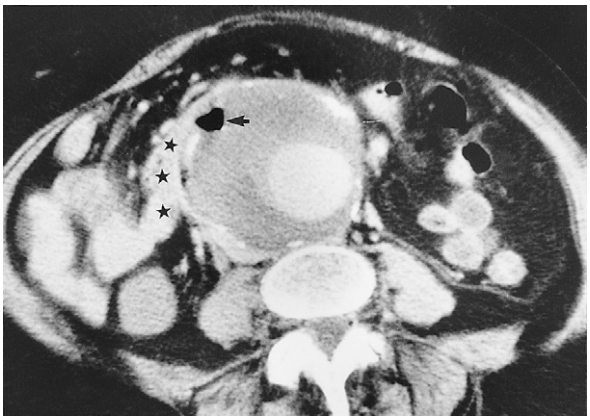
Barium meal A radiolucent mass within the duodenal cap with a thin coat of barium between the stone and duodenal wall



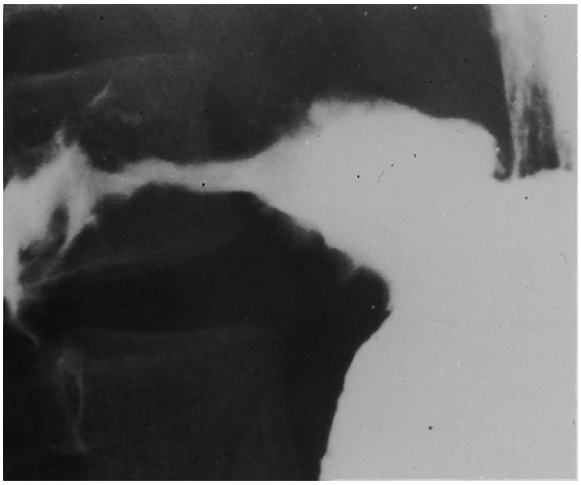
Superior mesenteric artery syndrome caused by carcinoma of the pancreas involving the root of the mesentery. (A) Supine position. Compression of the 3rd part of the duodenum. (B) Prone position. The compression persists and dilatation of the proximal duodenum is accentuated.†



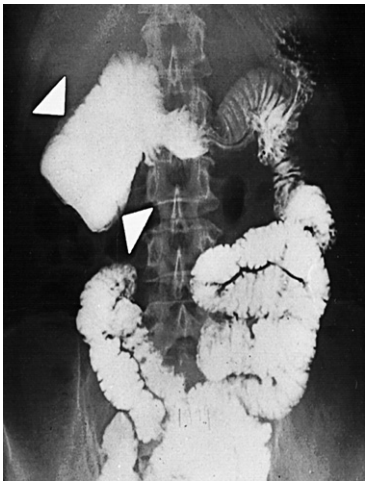
Intramural haematoma. CT shows a mass of mixed attenuation, characteristic of haematoma, surrounding the 3rd portion of the duodenum (arrowheads).*



Aortoduodenal fistula. Recent haematemesis. The 3rd part of the duodenum (stars) is stretched over the aortic aneurysm, which contains thrombus. A fistula accounts for the gas within the aortic wall (arrow).†



Crohn's disease. Marked irregular narrowing of the antrum and first portion of the duodenum, giving the 'pseudo post-Billroth I' appearance.*



Megaduodenum (arrowheads) in a patient with scleroderma.••

3.4 SMALL BOWEL

CROHN'S DISEASE

DEFINITION

- A chronic progressive *transmural* granulomatous inflammatory bowel disease
- There are typically discontinuous ('skip') lesions with asymmetrical bowel wall involvement
- It can affect any part of the GI tract – however it almost always affects the terminal ileum (in 95% of cases)

RADIOLOGICAL FEATURES

Barium studies

- **Bowel wall ulceration**
  - *Aphthoid ulcers:* characteristic superficial ulcers that do not penetrate the muscularis mucosa ► they appear as small collections of barium with surrounding radiolucent oedematous margins ► en face they appear as a dense amorphous barium pool with a surrounding black halo
    - They are typical of Crohn's disease (CD) (but are not seen with ulcerative colitis (UC))
  - *Fissuring 'rose thorn' ulcers:* deep ulcers with penetrating thorn-like cuts into the thickened intestinal wall ► they may lead to abscess formation, sinuses or fistulae
  - *Longitudinal ulcers:* these run along the ileal mesenteric border
  - *'Cobblestone' mucosa:* a combination of longitudinal and transverse ulceration separating intact portions of mucosa
- **Inflammatory polyps (pseudopolyps):** small, discrete round filling defects ► these are not a frequent finding
- **Thickened valvulae conniventes:** they can also be distorted, blunted or flattened (they are due to hyperplasia of the lymphoid tissue which causes an obstructive lymphoedema)
- **Bowel wall thickening**
  - Thickened bowel wall segments will displace adjacent barium-filled loops
  - Occasionally a smooth featureless outline will replace the normal mucosal pattern without a significant calibre change
  - *'Skip lesions':* discontinuous involvement of the bowel wall
  - *'Pseudodiverticula':* these are due to asymmetrical wall involvement and represent small patches of normal intestine in an otherwise severely involved segment
- **Strictures/stenoses**
  - These may be short, long, single or multiple (the latter is virtually diagnostic of CD) ► solitary strictures are common and may be accompanied by proximal (prestenotic) dilatation
  - *'String sign':* tubular narrowing of the intestinal lumen secondary to oedema and spasm (± scarring)
- **Fistulae formation:** this can involve adjacent loops of ileum, caecum or sigmoid colon
  - *Other sites:* urinary bladder ► perianal region (leading to a 'watering can' perineum) ► occasionally the skin and vagina
- **Bowel wall sacculation:** this is secondary to fibrosis within healing eccentric ulcers ► it can also be seen with ischaemic strictures or scleroderma (with wide 'square'-shaped diverticulae)

CT

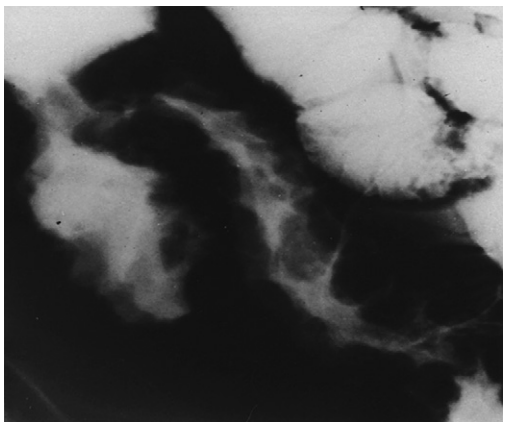
- **Thickened bowel wall**
  - The transmural disease leads to greater wall thickening than seen with UC ► mild reactive adenopathy (<1cm) can be present
  - *'Dirty fat':* transmural inflammation of the small bowel usually involves the adjacent mesentery
  - *'Target' or 'halo' sign:* a homogeneous or stratified appearance is seen on both NECT and CECT:
    - *NECT:* the stratified appearance is due to a thickened muscularis mucosae and submucosal fatty infiltration
    - *CECT:* the stratified appearance is due to acute inflammation with submucosal oedema and enhancement of the mucosa and muscularis propria
  - *Mesenteric fibrofatty proliferation:* this results in increased CT attenuation ► it is the most common cause of bowel loop separation in CD
  - *'Creeping fat':* fat accumulates on the serosal surfaces
  - *'Comb' sign:* mesenteric hypervascularity manifested as tortuosity, prominence and dilatation of the mesenteric arterial branches with a wide arrangement of the vasa recta
  - **Advanced disease:** intestinal perforation may lead to mesenteric phlegmon or interloop abscess formation ► these may contain gas – this is usually due to enteric or cutaneous fistulae and sinus tracts rather than due to a gas-producing bacteria

Magnetic resonance enterography/enteroclysis

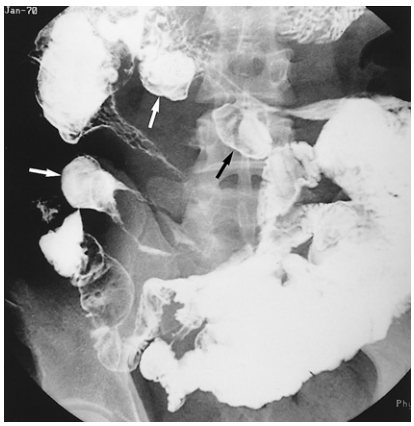
- This is an emerging technique which is able to assess disease activity and which is comparable with conventional enteroclysis (but it can also demonstrate extramural manifestations)
  - *It can demonstrate:* bowel wall thickening ► ulceration ► cobblestoning ► a 'target' appearance ► fibrofatty proliferation ► vascular engorgement ► adenopathy ► luminal narrowing and stenosis
  - *MR rectum:* this can assess any perianal fistulae (with fat suppressed T2WI and STIR sequences)

PEARLS

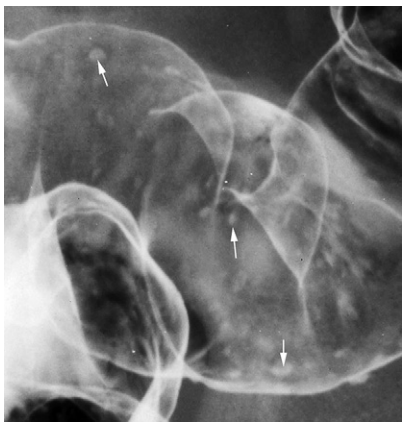
- **Complications:** there is an increased incidence of GI tract tumours, lymphoma and toxic megacolon
- **Extraintestinal findings:** gallstones ► spondylitis ► symmetrical sacroiliitis ► renal stones ► complications of steroid treatment (e.g. avascular necrosis) ► primary sclerosing cholangitis



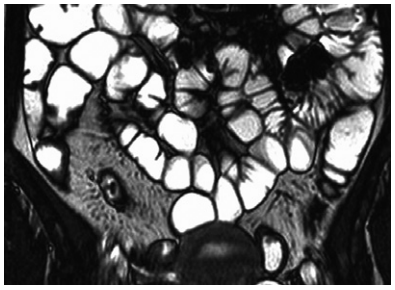
Cobblestoning of the terminal ileum, thickening of the wall of the terminal ileum, and an enlarged ileocaecal valve in Crohn's disease.*



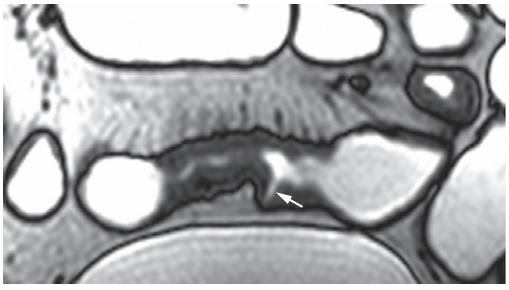
Advanced Crohn's disease with several characteristic pseudodiverticulae (arrows).†



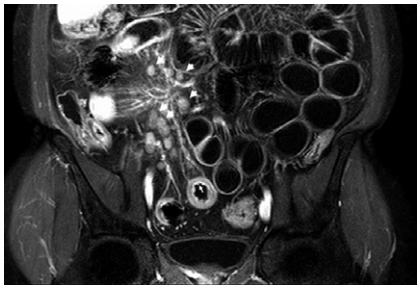
Aphthoid ulcers (arrows) in CD.*



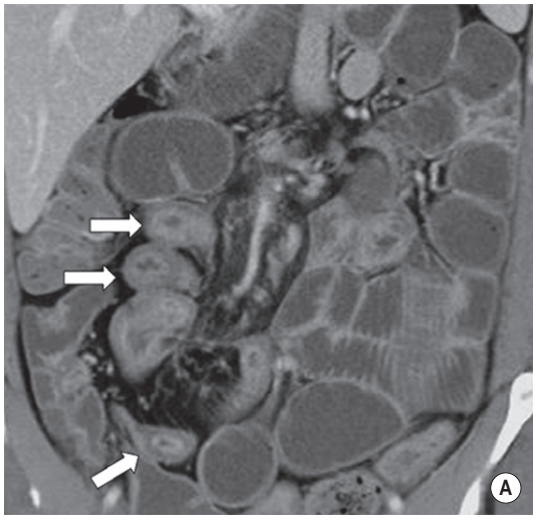
Extensive fibrofatty proliferation of the mesentery, accompanying involved ileal segments, is demonstrated on a coronal true FISP image.*



Active Crohn's disease. Coronal true FISP spot view demonstrates luminal narrowing and wall thickening in a segment of distal ileum. A fissure ulcer (arrow) penetrating the thickened wall and increased mesenteric vascularity are also disclosed.*



Active Crohn's disease. Coronal 2D FLASH image + Gad, showing multilayered mural enhancement of distal ileal loops (small arrow) and multiple enhancing mesenteric lymph nodes (arrowheads). Vascular engorgement is also present.*



Crohn's disease. (A) Coronal reconstruction image of CT enterography shows thickened distal ileal loops and mural stratification resulting in a 'target' appearance (arrows). Prestenotic dilatation is also seen. (B) A coronal, three-dimensional projection of the same patient showing the vascular engorgement (arrows) of an involved ileal loop (comb sign).*

TUBERCULOSIS (TB)

Definition Up to 5% of patients with *Mycobacterium tuberculosis* infection have GI involvement ► it used to be secondary to pulmonary disease but is now more likely to be of primary bovine origin (from drinking unpasteurized milk)

- *Affected sites:* GI tract ► liver ► spleen ► lymph nodes ► peritoneum ► female genital tract

Clinical presentation

- Abdominal pain ► fever ► weight loss ► diarrhoea ► intestinal obstruction ► bowel perforation (rare)

Radiological features

- Although any part of the GI tract can be involved, the ileum is the commonest involved site (esp. the terminal ileum and ileocaecal junction) ► there are often multiple bowel lesions
- Discrete transverse and circumferential ulcers, mucosal fold thickening and strictures are the main radiological features

Barium follow through (ileocaecal tuberculosis)

- Ulcerative, hypertrophic or fibrotic forms are described:
  - *Ulcerative:* there are discrete ulcers with a 'shaggy' edge – these tend to be large and circumferential
  - *Hypertrophic:* this presents as an inflammatory mass with associated bowel stenosis ► it may be difficult to distinguish from lymphoma
- **Fleischner sign:** a thickened patulous ileocaecal valve seen in conjunction with a narrowed terminal ileum
- **Stierlin's sign:** this is due to rapid emptying of contrast through a gaping incompetent ileocaecal valve and into a conically contracted caecum

CT Bowel wall thickening with homogeneous attenuation and lack of mural stratification ► there may be enlarged rim enhancing low-density mesenteric nodes (due to caseous liquefaction)

- *TB peritonitis:* this is suggested by diffuse omental and mesenteric infiltration, nodules, peritoneal thickening and high attenuation ascites

YERSINIA

Definition Infection with the Gram-negative bacilli *Yersinia enterocolitica* and *Yersinia pseudotuberculosis* ► acute inflammation of the terminal ileum is often indistinguishable clinically from an acute appendicitis

Barium follow through (BaFT) Disease is limited to the distal 20cm of ileum ► tortuous, thickened mucosal folds with small discrete nodular filling defects of lymphoid hyperplasia ► there can be mural thickening

ACTINOMYCOSIS

Definition Infection with *Actinomyces israelii*, a common saprophyte in the mouth, throat and GI tract ► the appendix is the most commonly affected site

- *Risk factors:* GI perforation ► previous surgery ► neoplasms ► diabetes ► steroids ► poor dental hygiene

BaFT and CT Appendix mass causing ileocaecal compression (± sinus tracts and fistulae)

GIARDIASIS

Definition A tropical infection with *Giardia lamblia* (a flagellate protozoon parasite of the upper small intestine) ► it is contracted through contaminated drinking water

BaFT Irregular thickening of the valvulae conniventes within the duodenum and proximal jejunum ► small well-defined nodular lymphoid hyperplasia in patients who also have dysgammaglobulinaemia

STRONGYLOIDIASIS

Definition A tropical small intestine infection with *Strongyloides stercoralis* (roundworm)

BaFT Delayed barium passage ► thickened or absent valvulae conniventes within the duodenum and proximal jejunum ► severe cases may show a rigid 'pipestem' stenosis with irregular narrowing

ANISAKIASIS

Definition Infection with *Anisakis* larvae (found within affected raw, pickled or salted fish): it can cause eosinophilic granuloma formation within the GI tract, affecting the stomach, terminal ileum

BaFT Concentric narrowing of the involved segment of ileum (± proximal dilatation) ► it may be indistinguishable from Crohn's disease (although the mucosa remains intact)

ASCARIASIS

Definition Infection with *Ascaris lumbricoides* (a roundworm) can cause intestinal or biliary obstruction, or oriental cholangiohepatitis ► it is very commonly seen in the tropics

BaFT Single or multiple smooth longitudinal or coiled filling defects within the small bowel (a thin central track of barium outlining the worm's intestinal tract may be seen)

CYTOMEGALOVIRUS

Definition Infection usually occurs in immunocompromised patients (especially AIDS)

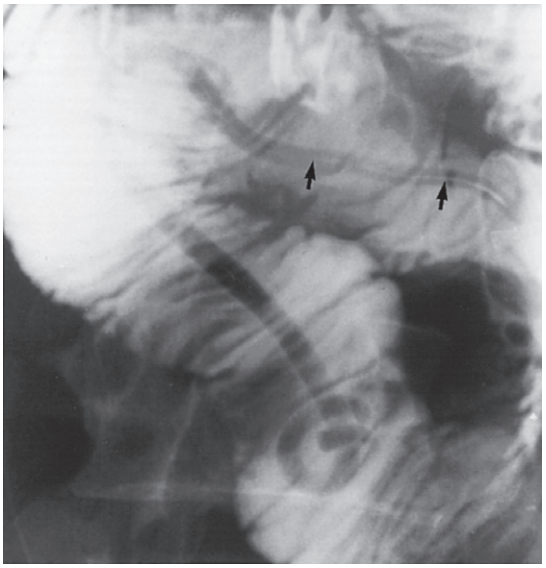
BaFT Diffuse small bowel narrowing, ulcerations and fold effacement ► there can be associated oesophagitis, gastritis or colitis

CRYPTOSPORIDIOSIS

Definition A protozoal infection causing enteritis in AIDS patients

BaFT Small bowel dilatation ► thickened small bowel folds

Giardiasis. Diffuse, irregular small bowel fold thickening is seen.••



Roundworm. Tubular intraluminal filling defect in the jejunum represents an ascaris worm. The thin white line bisecting part of the length of the worm (arrows) indicates barium in the worm's gastrointestinal tract.••

Tuberculosis. A short, irregular stricture, about 4cm long, is shown to involve the terminal ileum and ileocaecal valve. The narrowing caused considerable delay in the passage of barium, and dilatation of the ileum proximal to the stricture can be seen. The patient presented with intestinal obstruction.*

Colonic TB with thickening of the transverse colon (arrow) and extensive tuberculous ascites (asterisk).

CMV enteritis in a patient with AIDS. Tubular distal ileum (arrows) is seen.••

GASTROINTESTINAL STROMAL TUMOURS (GISTs)

Definition

- A submucosal mesenchymal (non-epithelial) tumour appearing to arise from the muscularis propria ► GISTs may arise from the interstitial cells of Cajal (these serve a gastric pacemaker function)
  - They are not related to a leiomyoma or leiomyosarcoma
- The commonest gastrointestinal mesenchymal tumour (< 1% of all GI tract tumours)
- *Location:* stomach (40–70%) > small intestine (20–50%) > large intestine and rectum (5%) ► leiomyomas and leiomyosarcomas are rare at these sites
  - *Oesophagus:* a leiomyoma is more common than a GIST

Clinical features

- It usually presents during the 6th to 7th decades (M = F)
- Asymptomatic (10–30%) ► abdominal pain ► abdominal mass ► ileus ► GI bleeding ► weight loss

Radiological features

- Malignant GISTs (10%) are radiographically indistinguishable from their benign counterparts (if >5cm, malignancy should be considered)

Barium meal Usually a small incidental and discrete submucosal mass (preserved overlying areae gastricae pattern) ► the border of the smooth mucosal surface forms right or obtuse angles with the adjacent mucosa

- Ulceration if it outstrips its blood supply (>2cm)
- *'Bulls- eye' or 'target' lesion:* this results from contrast collecting within the ulcer cavity

CT A well-circumscribed submucosal mass extending exophytically from the GI tract (± an intra- and extra-luminal 'dumb-bell' component) ► coarse mottled calcifications (25%) ► occasionally pedunculated (and may obstruct the pylorus or duodenum/act as a lead point for an intussusception) ► hypo- or hypervascular

- Lymphadenopathy is rare (its presence should suggest an alternative diagnosis such as lymphoma)
- The liver and peritoneum are the most common sites of distant metastases

MRI T1WI: intemediate SI ► T2WI: low to intermediate SI ► T1WI + Gad: variable heterogeneous enhancement

Pearls

- 90% of tumours will express KIT (CD 117) which is a tyrosine kinase growth factor receptor ► this differentiates a GIST from other gastrointestinal mesenchymal tumours (e.g. leiomyoma or leiomyosarcoma)
- Increased prevalence of GIST with NF-1
- *Unfavourable prognostic signs:* tumour size >5cm ► infiltration into adjacent organs ► metastases ► a high mitotic and proliferation index
- **Carney's triad:** a genetic syndrome of young women
  - Multiple stomach GISTs + a functioning extra-adrenal paraganglioma + a pulmonary chondroma

CARCINOID TUMOURS OF THE GASTROINTESTINAL TRACT

Definition

- A low-grade malignant neoplasm arising from submucosal neuroendocrine enterochomaffin cells
- It has a relatively indolent course with a prolonged survival (even with metastases)
- *Location:* appendix (45%) > small bowel (25%), colorectum (25%) ► it is multiple in ⅓ of cases

Clinical presentation

- The primary tumour rarely produces symptoms (due to its small size and deep mucosal location) ► it can present with abdominal pain, obstruction, or an abdominal mass ► GI haemorrhage is very rare
- **Carcinoid syndrome:** seen in ⅓ of jejunoileal carcinoids that have metastasized to the liver ► tumour secretion of serotonin can cause recurrent diarrhoea, bronchospasm, flushing, tricuspid insufficiency and pulmonary valvular stenosis
  - It can produce ACTH (Cushing's syndrome)

Radiological features

BaFT

- *Primary lesion:* a round, smoothly outlined intraluminal filling defect (it can be multiple)
- *Secondary mesenteric mass:* there is stretching, rigidity and fixation of the ileal loops
- Thickened valvulae conniventes (due to chronic ischaemic changes)
- A stellate, spoke-like arrangement of adjacent bowel loops or a sharp angulation of a bowel loop (due to the associated desmoplastic reaction)

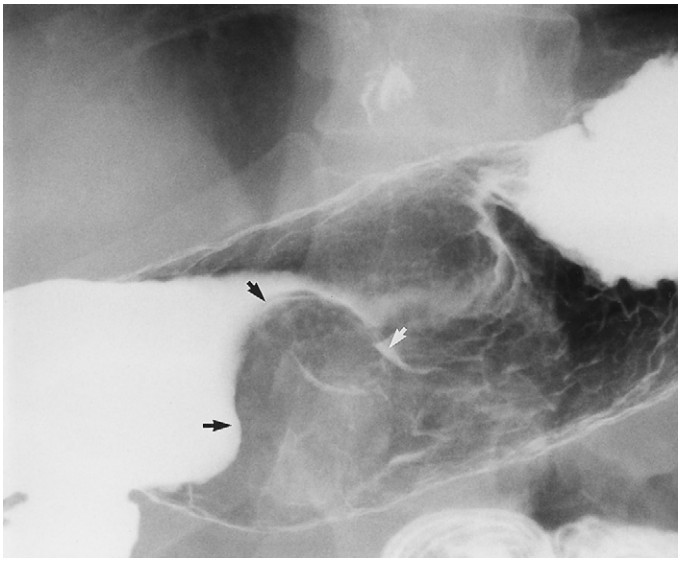
CT Secondary mesenteric changes including a discrete soft tissue mass (± stippled dystrophic calcification due to tumour necrosis) ► displaced adjacent bowel loops ► hypervascular liver metastases (which may calcify) ► mesenteric adenopathy (± dystrophic calcification)

- *Desmoplastic reaction:* this is incited by local serotonin release ► it can cause a radiating 'stellate' pattern of linear strands into the surrounding fat ► it may encase the mesenteric vessels with resultant chronic ischaemia and segmental bowel wall thickening

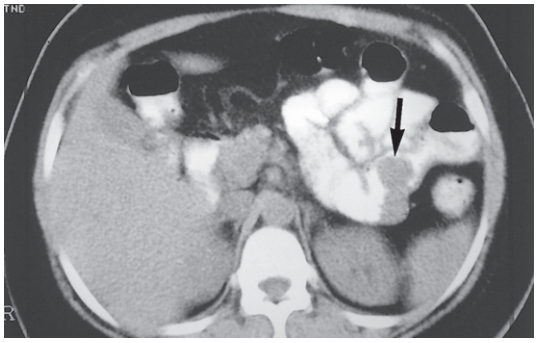
Pearls

- Small bowel carcinoids commonly metastasize ► gastric and appendiceal tumours rarely metastasize
- ^{123}I- or ^{131}I-MIBG scintigraphy can be used to localize and treat tumours

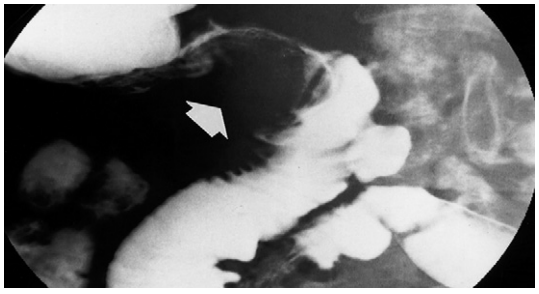
Malignant gastric stromal tumour. CT. This predominantly exophytic tumour is compressing the stomach (arrow).†



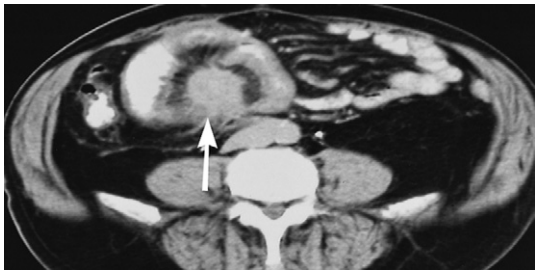
Benign GIST. The margins of this submucosal tumour make an obtuse angle with the adjacent normal mucosa.†



Benign stromal tumour. The tumour is visible on CT (arrow).†



Carcinoid tumour. A round, well-defined, intraluminal filling defect (arrow) is seen in the distal ileum of a patient who presented with symptoms of intermittent obstruction but without any manifestations of the carcinoid syndrome.†



CT shows a carcinoid mass (arrow) with a characteristic stellate radiating pattern and thickening of the adjacent intestinal wall.

Coronal reformat CECT showing a secondary mesenteric soft tissue mass containing dystrophic calcification and demonstrating surrounding desmoplastic reaction with radiating 'stellate' pattern of linear strands into the surrounding fat.

BENIGN TUMOURS

DEFINITION

- These arise from mucosa or submucosa (up to 2% of all GI tract neoplasms)
  - *Common:* adenoma (polyp or villous) ► leiomyoma – Only types with definite malignant potential
  - *Less common:* lipoma (3rd commonest tumour) ► a vascular or neurogenic tumour ► hamartoma (these are developmental anomalies – multiple lesions are seen in Peutz–Jeghers syndrome)

CLINICAL PRESENTATION

- Acute GI bleeding (commonly with a leiomyoma)
- Abdominal pain (commonly with a hamartoma causing a recurrent intussusception)

RADIOLOGICAL FEATURES

BaFT

- **Adenomatous polyp:** a small, smooth, intraluminal filling defect (often of different sizes and may be pedunculated) ► often solitary and sessile (if multiple it will usually affect a single segment)
- **Villous adenoma:** a broad-based, lobulated cauliflower-like filling defect with multiple radiolucent striations and frond-like projections ► usually >3cm in size
- **Leiomyoma:** these can lead to a tenting deformity of the intestinal wall, ulceration (± bleeding) and intussusception
  - *Intraluminal tumour:* a broad-based, smooth, round or semilunar filling defect
  - *Extraluminal tumour:* there is displacement and distortion of the neighbouring bowel loops
  - *Bidirectional or dumb-bell tumour:* both of the above

  CT A round or semilunar, homogeneous soft tissue mass associated with the bowel wall ► there is homogeneous or rim contrast enhancement (± peripheral crescent-shaped necrosis)

  DSA Well-defined, lobulated hypervascular mass
- **Hamartoma:** multiple round or lobulated filling defects ► often pedunculated (intussusception is frequent)
- **Lipoma:** a sharply marginated, solitary, sessile, intraluminal filling defect (3-4cm in size) ► it is easily deformed by peristalsis or compression on BaFT

  CT It will demonstrate fat attenuation

MALIGNANT TUMOURS

DEFINITION

- An adenocarcinoma is the most common malignant neoplasm of the small intestine ► it is usually solitary and located within the proximal small bowel

CLINICAL PRESENTATION

- It is almost always symptomatic (but with a non-specific presentation) ► there is a dismal prognosis due to the late diagnosis

RADIOLOGICAL FEATURES

Adenocarcinoma

BaFT An infiltrative tumour with circumferential narrowing, mucosal destruction and shouldering of its margins ► it can also appear as a filling defect or as a polypoid mass (± ulceration)

CT Mural thickening (not > 1.5cm) with either concentric or asymmetric luminal narrowing ► homogeneous or heterogeneous with moderate enhancement ► advanced tumours can infiltrate the mesentery ► lymphadenopathy (50%)

Carcinoid/lymphoma/malignant GIST: see Section 3 Chapter 4, Small bowel, Gastrointestinal stromal; Section 8 Chapter 2, Lymphoma

PEARLS

Secondary neoplasms arise by:

Direct invasion e.g. primary neoplasms of the ovary, colon, prostate, uterus and kidney

- These appear as a mass invading the adjacent intestine (often over a considerable length) ► there is mucosal destruction, luminal narrowing (± obstruction) and tethering of the mucosal folds

Lymphatic extension This is rare (e.g. spread of a caecal carcinoma to the terminal ileum)

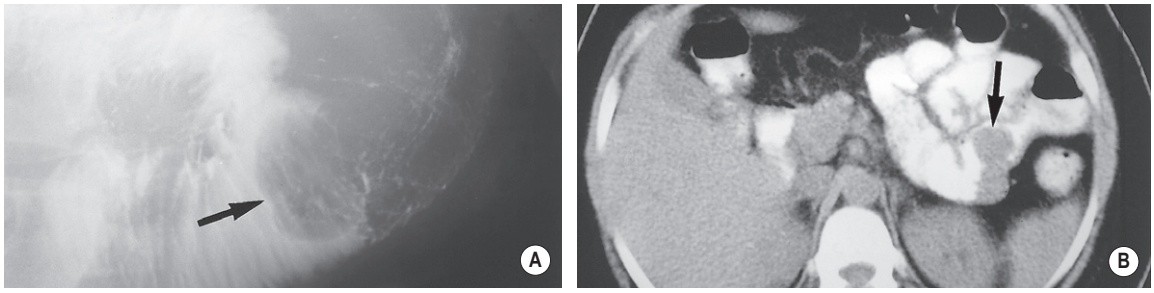
Peritoneal seeding These frequently localize within the right lower quadrant (stasis within the lower recesses of the distal mesentery allows deposition and growth)

CT Mesenteric infiltrates ► peritoneal implants ► omental 'caking' ► ileal loop separation with angled tethering of the mucosal folds

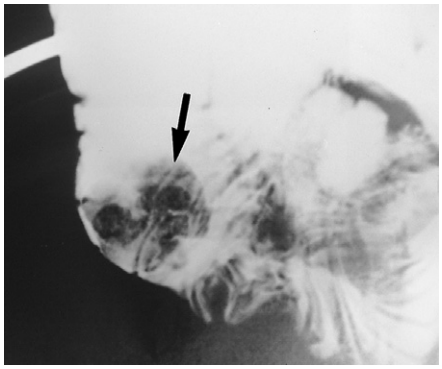
- *'Palisading':* the narrowed loops may align in a parallel configuration

Blood borne embolic metastases These are rare ► the common primaries are the lung, breast, kidneys and melanoma ► the resultant intraluminal soft tissue masses may lead to intussusception

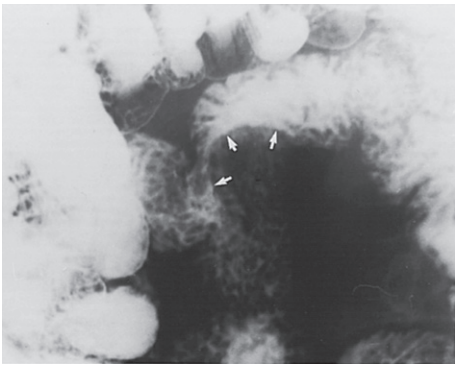
- Small, focal, nodular or infiltrating obstructive lesions
- Metastatic melanomas appear as multiple submucosal polypoid lesions ► the central ulceration gives a 'bull's eye' or 'target' appearance on a BaFT



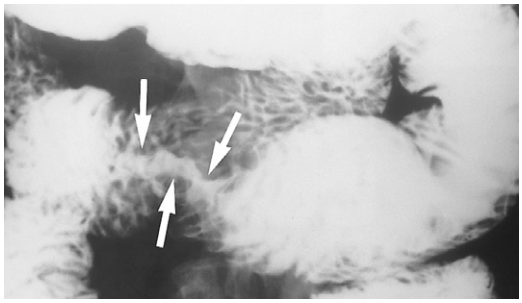
Benign stromal tumour. (A) BaFT reveals an intraluminal mass (arrow) on compression. (B) The tumour is also visible on CT (arrow).†



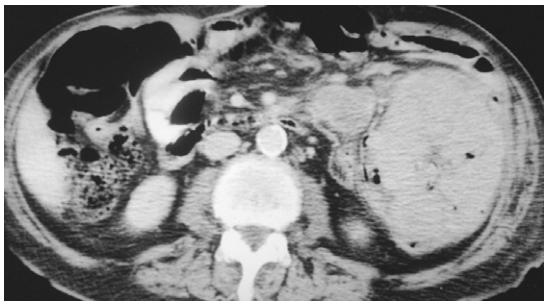
BaFT reveals an ileal hamartoma (arrow) in Peutz–Jeghers syndrome.



Submucosal mass (arrows) represents a jejunal leiomyoma.••

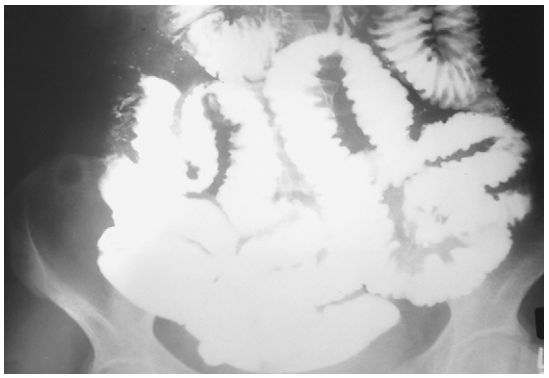


Small bowel adenocarcinoma (between arrows).†



Lymphoma. CT reveals a well-demarcated soft tissue mass.†

Carcinoid tumour. A round, well-defined, intraluminal filling defect (arrow) is seen in the distal ileum of a patient who presented with symptoms of intermittent obstruction but without any manifestations of the carcinoid syndrome.*



Lymphoma. Diffuse fold thickening and nodularity.†

MECHANICAL SMALL BOWEL OBSTRUCTION

Causes

- **Mural lesions:** tumour ► Crohn's stricture ► irradiation ► ischaemia
- **Luminal lesions:** bezoar ► gallstone ► *Ascaris lumbricoides* bolus ► intussusception
- **Extrinsic lesions:** volvulus ► abdominal malignancy
  - *Adhesions:* this accounts for 75% of cases in developed countries
  - *Hernias:* this accounts for 75% of cases in underdeveloped countries

Radiological features

Erect AXR This is not routinely performed ► >2 fluid levels within dilated (> 2.5cm) small bowel loops

Supine AXR Small bowel dilatation (this can be gas or fluid filled – fluid-filled loops are not easily seen) ► collapsed colon ► a cause may be identified (e.g. an inguinal hernia may appear as a gas-filled viscus below the level of the inguinal ligament)

- *'String of beads' sign:* a line of gas bubbles trapped between the valvulae conniventes within almost completely fluid-filled and very dilated small bowel

US This can detect fluid-filled loops of small bowel but is rarely used

CT Adhesions are suggested by angulated and tethering of the bowel loops ► a cause may be identified (e.g. neoplasm)

- *Simple obstruction:* a transition zone may be seen with dilated small bowel loops proximal to the obstruction, and collapsed loops distally
- *Closed-loop obstruction:* a U- or V-shaped configuration of the dilated loops with a fixed radial distribution
- *Strangulated bowel:* this represents incarceration of the two limbs of the mechanical small bowel obstruction with subsequent ischaemia

CT signs*

Closed-loop intestinal obstruction	Strangulating obstruction
Dilated fluid-filled loops	Wall thickening of affected loop
U-shape configuration	High attenuation* in bowel wall
Thickening of mesenteric vessels	Gas in bowel wall
Radial spread of mesenteric vessels	Gas in mesenteric veins
Tapering of the loop ('beak' sign)	Mesenteric congestion
Triangular loop	Mesenteric haemorrhage
Twisted mesentery ('whirl' sign)	Poor or no contrast enhancement

*Representing haemorrhage

INTUSSUSCEPTION

Definition

- Telescoping of a segment of proximal bowel into a segment of distal bowel
  - *Intussuscipiens:* the part of bowel into which another part is prolapsed
  - *Intussusceptum:* the part of bowel that has prolapsed
- **Children:** a common surgical emergency a (peak incidence between 5 and 9 months of age) ► presents with intermittent colicky abdominal pain and 'redcurrant jelly' stools ► usually ileocolic (it can also be ileo-ileocolic, ileo-ileal and colocolic) ► >90% of children have no demonstrable lead point (the cause is often lymphoid hypertrophy)
  - *Other causes*: Meckel's diverticulum ► intestinal polyp ► duplication cyst ► lymphoma
- **Adults:** it is nearly always caused by a bowel neoplasm ► a colonic lipoma, lymphoma and melanoma metastases are other causes

Radiological features

AXR A soft tissue mass (possibly part-outlined by gas) ► if orientated end-on a target sign may be seen (consisting of two concentric circles of fat density alternating with soft tissue density) ► small bowel obstruction

US A 'pseudotumour' or 'kidney' sign

CT The intussusceptum brings the mesenteric fat into the lumen of the intussuscipiens ► an intussusception appears as a sausage-shaped mass or as a 'target' mass (depending on its orientation)

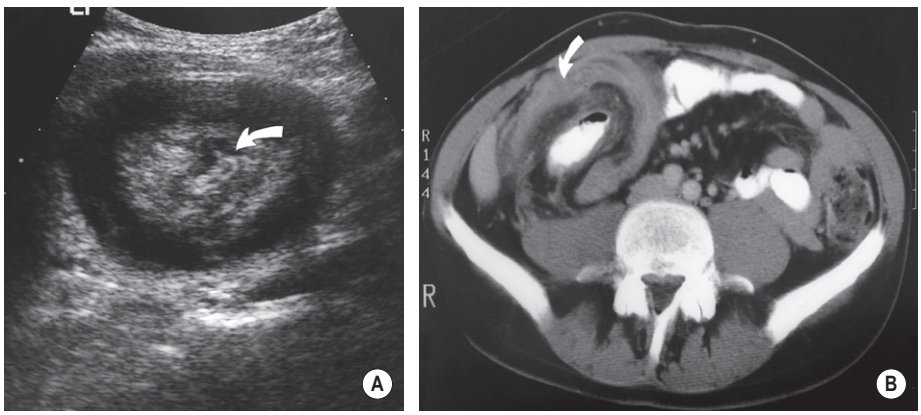
Pearls

- **Treatment:** pneumatic air reduction under fluoroscopic guidance or hydrostatic reduction under US control ► this is contraindicated if there is free gas present, septicaemic shock or peritonitis
- **Pneumatic reduction:** this should only use a maximum pressure of 120mmHg
  - 3 attempts for 3 minutes is recommended (initially at a pressure of 60–80mmHg)

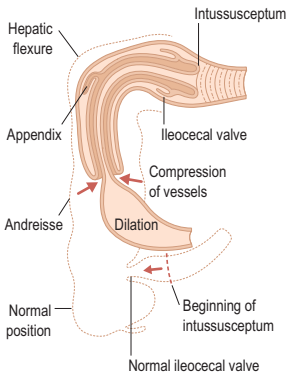
Small bowel obstruction. Supine AXR showing very distended small bowel identified by its central position, multiple loops and valvulae conniventes.*

Adhesive small bowel obstruction. Coronal reformat CECT shows the transition zone (white arrow), distended, fluid-filled proximal jejunal loops and collapsed distal loops (black arrow). No mass is seen. *If the bowel becomes oedematous (e.g. ischaemia) these folds may become thickened and difficult to distinguish from colonic haustra.*

Distiniguishing features of small and large bowel dilatation		
	Small bowel dilatation	**Large bowel dilatation**
Number of bowel loops	Usually numerous	Fewer loops
Distribution of bowel	Central abdomen	Peripheral abdomen
Size of bowel	Rarely > 5 cm	Often > 5 cm
Fold pattern	Valvulae conniventes: thin complete bands across the small bowel ► the folds are closer together than colonic haustra ► they are most prominent in the jejunum	Colonic haustra: these are usually thick incomplete bands ► they may be absent from the descending and sigmoid colon
Bowel contents	Fluid and gas	Faeces and gas



Ileocolic lymphoma leading to ileocolic intussusception. (A) US image of the right iliac fossa showing the 'pseudotumour' or 'kidney' sign. The ileum can be seen centrally (arrow), surrounded by mesenteric fat that is hyperechoic, all within the thickened ascending colon. (B) CT showing oral contrast medium in the ileal lumen, the surrounding mesenteric fat accompanying the intussusceptum and the thickened ascending colon, which is the intussuscipiens (arrow).*



Schematic representation of an ileocolic intussusception.

JEJUNAL DIVERTICULA

Definition

- Mucosal herniation through the jejunal wall ► these are uncommon
  - *Ileal diverticula:* these are rarer still (affecting the mesenteric border of the terminal ileum) ► they are also smaller and fewer in number

Clinical presentation

- The 'blind loop' syndrome (due to bacterial overgrowth): abdominal pain + distension + weight loss + a megaloblastic anaemia
  - *Less common complications:* acute diverticulitis ± perforation ► a mesenteric abscess ► bleeding or small bowel obstruction

BaFT A relatively large narrow-necked outpouching affecting the mesenteric border ► they are usually multiple

MECKEL'S DIVERTICULUM

Definition

- An ileal outpouching following failure of the yolk sac to close during fetal life ► they are found in up to 3% of the population
  - *Location:* the antimesenteric border of the ileum (30–90cm from the ileocaecal valve and measuring between 0.5 and 15cm)
  - Ectopic gastric mucosa can be found within a diverticulum in 20% of adults and 95% of children presenting with bleeding
  - *Complications:* ulceration ► bleeding ► perforation ► inflammation ► intussusception ► internal herniation ► volvulus ► adhesions

^{99m}Tc-pertechnetate Increased uptake if gastric mucosa is present ► this is more accurate in children

Enteroclysis A blind-ending sac arising from the antimesenteric border of the ileum ► a triradiate pattern of mucosal folds may be seem at the diverticulum base

Angiography It can demonstrate a persistent vitelline artery in patients presenting with chronic bleeding

WHIPPLE'S DISEASE

Definition

- A rare chronic bacterial infection with *Tropheryma whippelii* (a Gram-positive bacillus) causing abdominal pain, diarrhoea, malabsorption, adenopathy and polyarthritis

BaFT Thickened valvulae conniventes (often with a micronodular appearance within the proximal small intestine)

CT Non-specific bowel wall thickening ► low-density retroperitoneal and mesenteric lymphadenopathy (due to an increased amount of fat and fatty acids)

SMALL BOWEL FISTULAE

Definition

- These are associated with Crohn's disease, diverticulitis, malignancy (e.g. colorectal cancer), and in the postoperative patient
  - *Enterocolonic:* small bowel to colon
  - *Enteroenteric:* small bowel to small bowel
  - *Enterocutaneous:* small bowel to skin
  - *Enterovaginal:* small bowel to vagina
  - *Enterovesical:* small bowel to bladder

Fistulogram This allows assessment via the use of water-soluble contrast injected into the fistula (using a small catheter)

- *Alternative methods of diagnosis:* a small bowel FT or barium enema ► CT or MRI (with oral contrast medium)

ACUTE MESENTERIC ISCHAEMIA

Definition

- A compromised small intestinal blood supply due to mesenteric arterial embolism or thrombosis, mesenteric venous occlusion or low flow states

Clinical presentation

- Severe abdominal pain (which can be out of proportion to the clinical signs) ► a lactic acidosis (due to infarcted tissue)

CT Dual-phase imaging with oral (water) and IV contrast medium is required for accurate mesenteric vessel evaluation

- *Thrombus:* a filling defect within a mesenteric artery or vein
- *Affected bowel loops:* bowel distension ► circumferential bowel wall thickening (with hypoattenuation due to submucosal oedema) ► reduced, delayed or absent bowel wall enhancement ► increased mesenteric fat attenuation ► engorged mesenteric veins ► ascites
  - *Late signs:* pneumatosis ► mesenteric or portal venous gas represents irreversible ischaemia

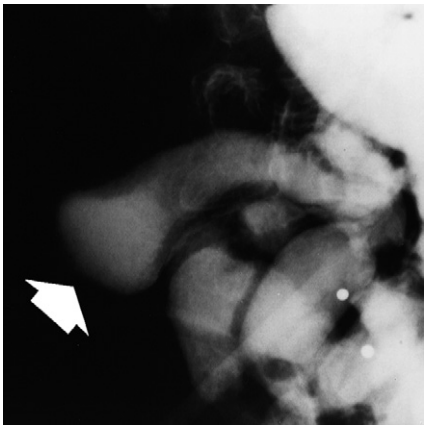
NODULAR LYMPHOID HYPERPLASIA

Definition

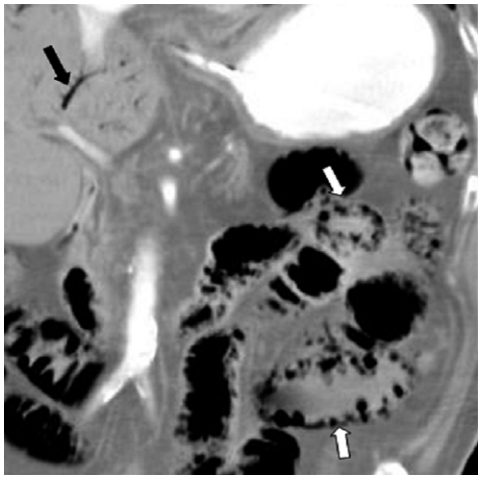
- This is a normal finding within the terminal ileum in children or young adults
  - In older adults it is associated with immunoglobulin deficiency (particularly late-onset hypogammaglobulinaemia)

BaFT Multiple small (1–3mm) discrete round lesions throughout the small intestine (increasing in number as one travels distally) ► the colon is frequently involved throughout its length

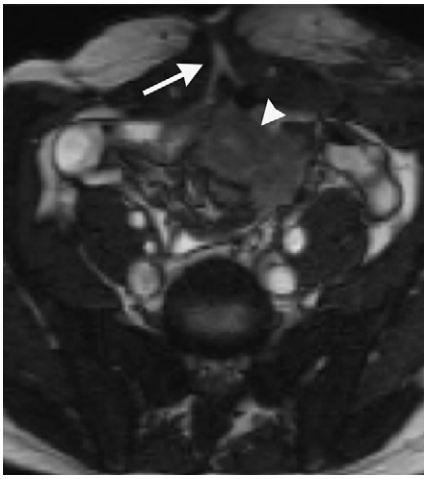
Mesenteric venous thrombosis. CECT, coronal reformation image shows circumferential thickening of distended jejunal loops (white arrows) and haziness of the adjacent mesentery. Non-opacification of superior mesenteric and jejunal branches is noted (black arrows). Ascites is also present.*



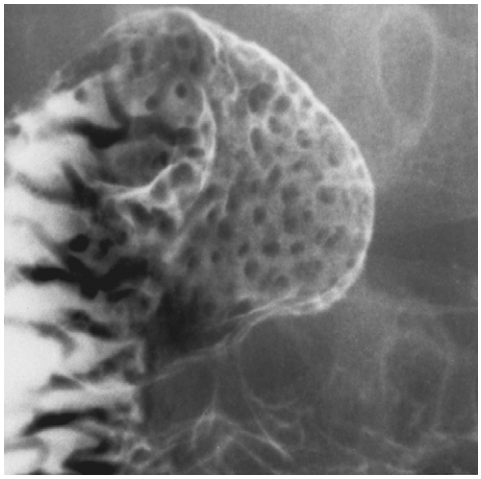
Meckel's diverticulum. Follow through study demonstrating a blind-ending sac is shown arising from the antimesenteric border of the distal ileum (arrow).*



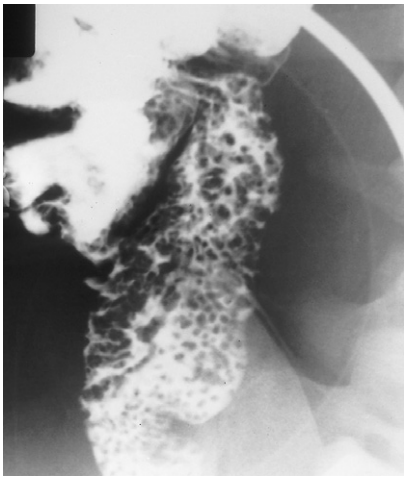
Small bowel infarction. CECT shows pneumatosis of small bowel loops (white arrows) and hepatic portal vein gas (black arrow).



Enterocutaneous fistula. Axial true FISP showing a high SI fistula extending from an inflamed loop of small bowel (arrowhead) to skin (arrow) in a patient with Crohn's disease.*



Lymphoid hyperplasia. Multiple small filling defects characteristic of lymphoid hyperplasia are shown on a double-contrast view of the duodenal cap.*



Follow through study demonstrating terminal ileum nodular lymphoid hyperplasia.

COELIAC DISEASE (NON-TROPICAL SPRUE/GLUTEN-SENSITIVE ENTEROPATHY)

Definition

- *Non-tropical sprue (coeliac disease):* a disorder of the small intestinal mucosa caused by intolerance to α gliaden (a component of gluten) ► it tends to affect the more proximal small bowel
  - Genetically susceptible individuals are usually children or young adults
- *Tropical sprue:* a malabsorption state seen within tropical countries and which affects the entire small bowel ► there is a possible infective aetiology (it is distinct from coeliac disease)

Clinical presentation

- Symptoms are secondary to malabsorption: diarrhoea ► weight loss ► steatorrhoea ► malnutrition ► anaemia ► abdominal pain

BaFT Dilated bowel loops ► straightened and thickened jejunal valvulae conniventes

- *Flocculation:* a coarse appearance of small clumps of disintegrated barium (due to the increased intestinal fluid)
- *Segmentation:* of a normally continuous barium column
- *'Moulage' sign:* mucosal atrophy and absence of valvulae
- *'Jejunization' of the ileum:* the presence of numerous mucosal folds within the ileum (reversal of the normal jejunoilieal fold pattern)
- *'Colied spring' appearance:* due to a transient non-obstructive intussusception

CT As above but also: bowel wall thickening ► small volume ascites ► vascular engorgement ► low attenuation mesenteric lymphadenopathy ► cavitating mesenteric lymph nodes

Pearls

- **Diagnosis:** this is based upon an abnormal villous pattern detected with a peroral jejunal biopsy (radiological investigation is generally reserved for those with a normal biopsy or suspected complications)
- **Associated disorders:** dermatitis herpetiformis ► IgA deficiency ► hyposplenism
- **Complications:** there is an increased risk of GI T-cell lymphoma or oesophageal and jejunal carcinoma
  - *Ulcerative jejunoileitis:* segments of bowel wall thickening with irregularity and ulceration

GALLSTONE ILEUS

Definition

- A rare condition caused by a gallstone eroding through an inflamed gallbladder and passing into the adjacent duodenum – this will usually pass distally until it impacts at the narrowed terminal ileum (causing obstruction)
  - *Bouveret's syndrome:* the gallstone passes proximally into the stomach

AXR/CT Small bowel obstruction ► an obstructing gallstone within the pelvis ► gas within the biliary tree (due to retrograde passage of air from the duodenum through the fistula)

- *Gas within the liver parenchyma:* biliary gas tends to normally be centrally located (portal venous gas will tend to have a more peripheral distribution)

PROGRESSIVE SYSTEMIC SCLEROSIS (SCLERODERMA)

Definition

- A collagen vascular disease of unknown aetiology – smooth muscle atrophy is followed by collagen deposition and fibrosis ► it affects the skin, joints, blood vessels and viscera

BaFT A dilated duodenum and jejunum ► reduced peristalsis with an increased transit time ► pneumatosis intestinalis

- *Sacculations (pseudodiverticula):* large broad-based outpouchings with a squared contour seen on the antimesenteric small bowel border
- *'Hidebound' appearance:* an increased number of mucosal folds

Associations A dilated oesophagus and reflux oesophagitis (± stricture) ► soft tissue calcification ► pulmonary interstitial fibrosis ► acro-osteolysis

SMALL BOWEL DISORDERS DUE TO CELLULAR INFILTRATION: EOSINOPHILIC GASTROENTERITIS

Definition

- Eosinophilic infiltration of the walls of the stomach and small intestine

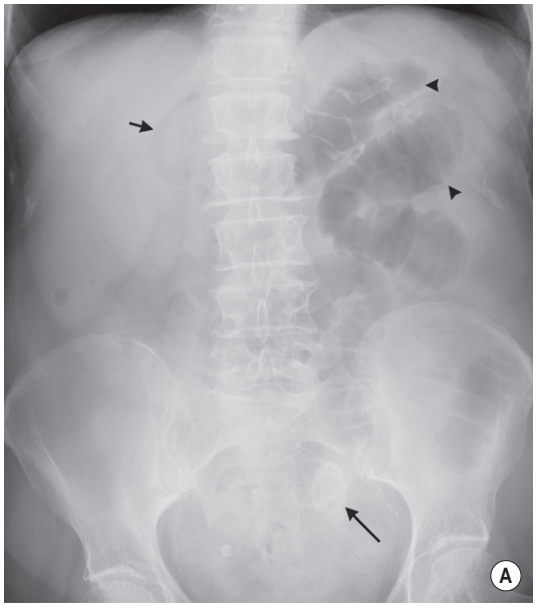
BaFT Thickened valvulae conniventes and mural thickening (± bowel obstruction) ► gastric nodularity ► a narrowed pyloric antrum

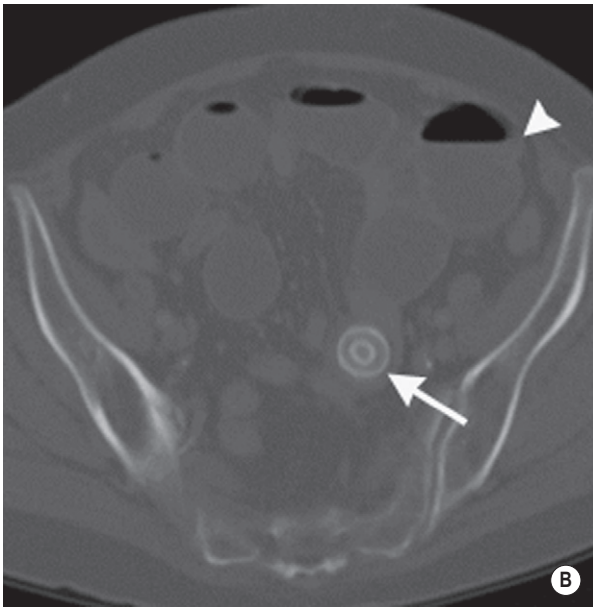
SMALL BOWEL DISORDERS DUE TO CELLULAR INFILTRATION: MASTOCYTOSIS

Definition

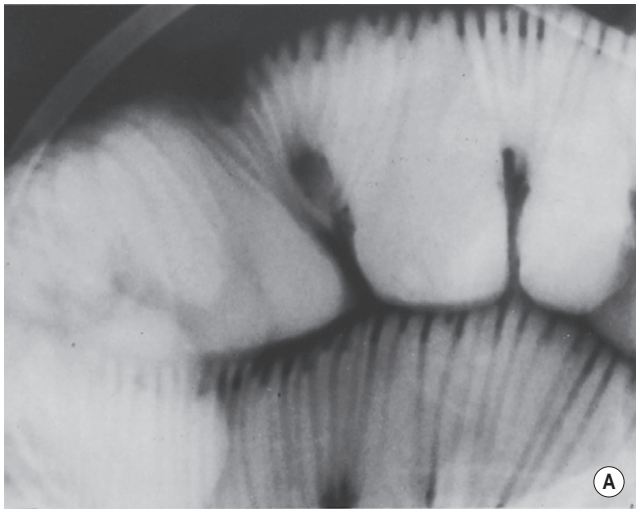
- This is due to mast cell infiltration of the small intestine ► skin infiltration causes a typical skin rash (urticaria pigmentosa)
  - It is a associated with hepatomegaly, dense bones and peptic ulcer disease

BaFT Thickened valvulae conniventes ► 2–5mm nodular mucosal defects – these are usually seen within short jejunal segments (they can also be seen within the ileum)

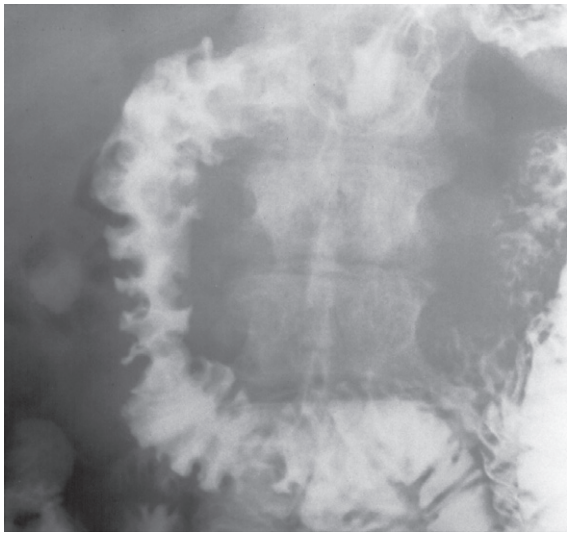




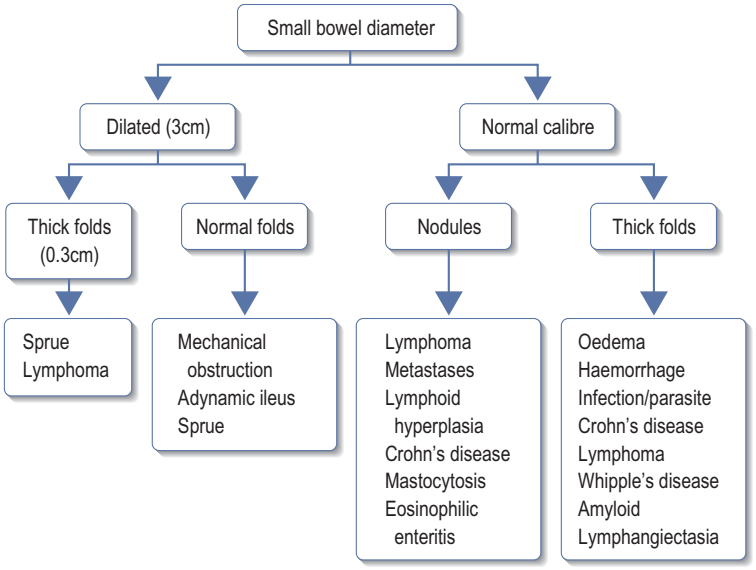
(A) Plain AXR demonstrating a pelvic gallstone (long arrow), small bowel obstruction (arrowheads) and gas within the biliary tree (short arrow). (B) CT confirming the laminated pelvic gallstone (arrow) and small bowel obstruction (arrowhead).



Systemic sclerosis. (A) Sacculation shown as broad-based outpouchings. (B) Dilatation of a segment of intestine with the 'hide-bound' appearance of the valvulae conniventes seen on a spot compression view.*



Mastocytosis. Nodular asymmetrical fold thickening of the duodenum is seen. Note sclerotic bones.••



Basic algorithm for small bowel disease. Many diseases have a variable appearance.

INTESTINAL LYMPHANGIECTASIA

Definition

- *Primary disease:* part of a generalized lymphatic channel hypoplasia (with generalized lymphoedema, chylous pleural effusions, malabsorption and lymphocytopenia) which is seen in children or young adults
- *Secondary disease:* lymph flow is obstructed by retroperitoneal fibrosis or malignant infiltration of the mesenteric and retroperitoneal lymph nodes

Radiological features

BaFT Non-specific uniformly thickened, closely set and parallel valvulae conniventes (± a micronodular mucosal surface pattern)

CT Mural thickening

AIDS

Definition

- The small bowel is affected in 50% of patients by opportunistic infections (commonly *M. avium-intracellulare* and cryptosporidiosis), Kaposi's sarcoma and AIDS-related lymphoma

Radiological features

BaFT Thickened (± nodular) valvulae conniventes within the proximal small intestine

CT Bulky retroperitoneal and mesenteric nodal masses are seen that are often indistinguishable from Kaposi's sarcoma or lymphoma

GRAFT-VERSUS-HOST DISEASE

Definition

- This develops following an allogenic bone marrow transplantation – the foreign donor lymphoid graft tissue mounts an immunological reaction against the hosts skin, liver and GI tract

Radiological features

BaFT

- *Acute phase (4–15 days):* uniform thickening or flattening of the mucosal folds ► a thickened intestinal wall ► ribbon-like luminal narrowing and ulceration throughout the jejunum and ileum
- *Subacute phase (13–96 days):* this is similar to the acute phase (often with a striking segmental distribution)
- *Resolution phase:* improvement is demonstrated (with no mucosal fold abnormality) but there is mural thickening confined to the terminal ileum

CHRONIC RADIATION ENTERITIS

Definition

- Intestinal ischaemia secondary to previous radiotherapy (with a possible lag time of 25 years) ► it results in damage to the vascular endothelial cells which leads to an endarteritis obliterans ► the distal ileum (esp. its pelvic loops) is the most frequently affected region
  - *Risk factors:* high radiation doses ► radiation treatment over a short time ► a large treatment volume

Clinical presentation

- Colicky abdominal pain ► diarrhoea ► malabsorption ► intermittent small intestinal obstruction

Radiological features

BaFT Thickened valvulae conniventes ► mural thickening ► effacement of the mucosal pattern ► ulceration, fixation and angulation of the small intestinal loops ► luminal narrowing and stenosis ► sinus and fistulae formation (uncommon) ► linear streaks of increased attenuation within the mesenteric fat (secondary to oedema)

- *'Mucosal tacking':* spiking and distortion of antimesenteric mucosal folds (caused by adhesions to inflamed and thickened mesentery) ► mesenteric retraction

CT This is best for assessing any mural thickening

- *Acute phase:* a target configuration (due to oedema and inflammation)
- *Chronic healing fibrotic phase:* homogeneous mural thickening

NON-STEROIDAL ANTI-INFLAMMATORY DRUGS (NSAIDs)

Definition

- Patients may develop non-specific small intestinal ulceration (with blood and protein loss) on long-term treatment

Radiological features

BaFT Concentric, circumferential diaphragm-like narrowings (submucosal fibrosis secondary to focal ulceration) ► this may progress to stricture formation

AMYLOIDOSIS

Definition

- Infiltration of the GI tract with amyloid occurs in the majority of patients with primary amyloidosis

Radiological features

BaFT Amyloid deposition can cause symmetrical thickening or effacement of the valvulae conniventes (as well as atrophy) ► intraluminal masses of amyloid ► bowel dilatation

CT Non-specific symmetric wall thickening

BEHÇET'S DISEASE

Definition

- A chronic multisystem vasculitis involving the mucocutaneous, ocular, cardiovascular, gastrointestinal and central nervous systems

Radiological features

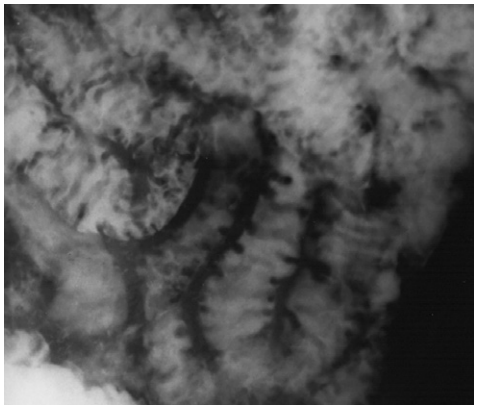
- It resembles ileocaecal tuberculosis or Crohn's disease

BaFT Deep discrete ulceration leading to haemorrhage or perforation

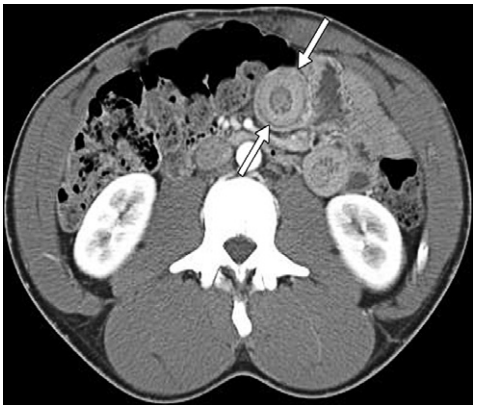
CT A polypoid mucosa with wall thickening and marked enhancement ► there is little adenopathy, fibrofatty change or pericolonic inflammation unless perforation has occurred (cf. Crohn's)

Graft-versus-host disease. Diffuse effacement of small and large bowel folds are seen. Note the ribbon-like appearance of small bowel.••

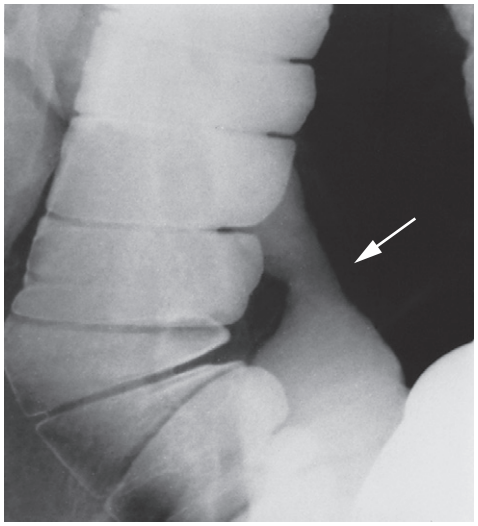
M. avium-intracellulare in a patient with AIDS. Diffuse small bowel fold thickening is seen.••



Lymphangiectasia. Diffuse small bowel fold thickening is seen.••



Patient with AIDS and involvement of the small bowel. Note the thickened wall (arrows).••



Radiation stricture. This spot view of a barium infusion examination shows a short, tight stricture of the terminal ileum in a patient who 4 years earlier had undergone radiotherapy for carcinoma of the uterus.*

BaFT in a patient with extensive radiation enteritis reveals strictures, dilatation and a 'picket-fence' appearance (arrows).†

3.5 COLON

ULCERATIVE COLITIS (UC)

Definition

- A relapsing and remitting inflammatory bowel disease predominantly involving the colorectal mucosa and submucosa ► there is symmetrical colonic involvement (cf. asymmetrical Crohn's disease)
- It always involves the rectum – any remaining colitis is in continuity with its proximal extent
  - *Proctitis:* inflammatory changes limited to the rectum

Clinical presentation

- Bloody diarrhoea (± constitutional symptoms) in young adults ► an acute fulminating colitis with a risk of perforation (15%) ► extracolonic manifestations:
  - Synovitis ► ankylosing spondylitis ► sacroiliitis ► erythema nodosum ► pyoderma gangrenosum ► primary sclerosing cholangitis ► cholangiocarcinoma ► iritis

Radiological features

Double-contrast barium enema (DCBE)

- **Crypt abscesses:** may erode through the muscularis mucosae and spread laterally within the submucosa:
  - *En face appearance:* linear, transverse, serpiginous or rounded
  - *Tangential appearance:* undercutting of the mucosal edge can give a 'T' or 'collar stud' shape
- Ruptured crypt abscesses lead to superficial erosions which fill with barium to produce a typical granular mucosal pattern (producing continuous ulceration on a background of diffusely abnormal mucosa – discrete ulceration with normal intervening mucosa is not seen)
- **Reflux ileitis:** there is a patulous ileocaecal valve and a granular distal ileum
- **Postinflammatory polyps:** when an acute attack remits, the granulation tissue forming at the ulcer base undermines the residual oedematous mucosal flap at the ulcer edge – this is therefore prevented from sealing down, resulting in sessile, filiform, frond-like polyps (less commonly found in Crohn's disease)
- **Chronic colitis:** a tubular, shortened, featureless ('lead-pipe') colon
- **Strictures:** chronic hypertrophy of the muscularis mucosa (and submucosal thickening with fat) can cause generalized colonic shortening as well as localized left-sided colonic strictures (10–20%) ► the strictures are smooth, tapering and symmetrical (cf. asymmetrical strictures in Crohn's disease)

US

- Wall thickening (≥ 4mm) ► a stratified appearance with differentiation between the submucosa and muscularis propria ► ulceration (with focal disruption of the bowel wall layers which may be outlined by intracolonic gas) ► inflammatory echogenic pericolic fat

CT

- *Acute disease:* wall thickening (≥ 4mm) tending to be less marked than with Crohn's disease ► absence of formed faecal residue within any affected segments ► normal pericolonic tissues (unless perforation has occurred)
- *Chronic disease:* a widened presacral space due to fibrofatty proliferation
- *'Target' sign:* due to chronic muscularis mucosae thickening and fatty submucosal infiltration (visible even with NECT)

Pearls

- There is an increased risk of colorectal carcinoma (due to dysplastic changes within diseased epithelium rather than from a prior adenoma) – this is more common with an extensive colitis of > 10 years duration ► tumours are frequently multiple and infiltrative
  - *Dysplasia-associated lesions (DALMs):* representing severe dysplasia and are a very high risk marker for cancer (similar to a villous adenoma)
  - *Early infiltrative carcinoma:* this presents with a fixed, irregular, in-drawn base
  - *Strictures:* these are usually benign ► malignancy is suggested by an irregular raised area, shouldering or asymmetry
- **Complications:** toxic megacolon ► perforation

TOXIC MEGACOLON

Definition

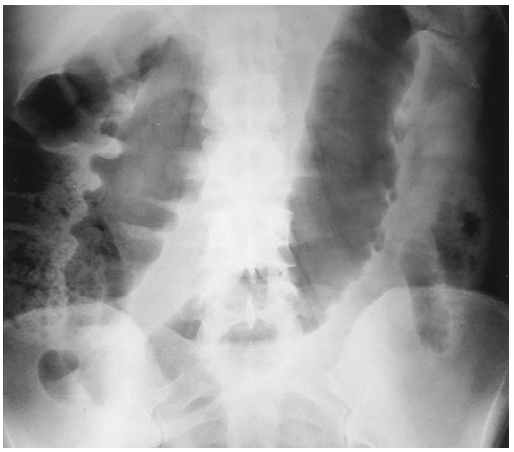
- A fulminating colitis: transmural inflammation and ulceration extends deep into the muscular layers with neuromuscular degeneration ► it accounts for most UC-related deaths
- It can also occur in any other cause of colitis but is less frequently seen in Crohn's disease, bacterial colitis, pseudomembranous colitis or ischaemic colitis

Radiological features

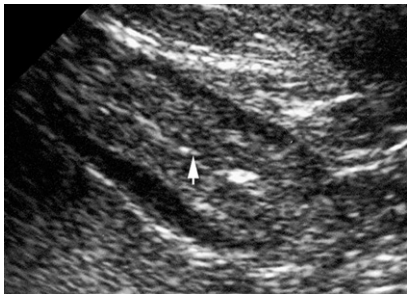
- It usually affects the transverse colon (the least dependent part of the colon where intraluminal gas collects) ► perforation is frequent
  - **Dilatation:** if > 5cm it is associated with deep ulceration into the muscular layers (> 8.5cm in established cases) ► the haustra are always absent
- A daily plain AXR is important for assessing and monitoring the colitis extent (barium studies are contraindicated due to the perforation risk)

Chronic UC with mesorectal lipohyperplasia causing widening of the post-rectal space. There is increased submucosal fat (arrow) creating a target sign in this NECT.*

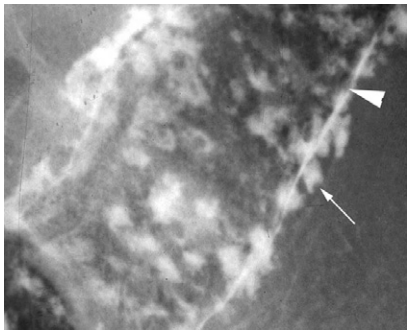
The granular mucosa typical of UC. Note the intact mucosal line.*



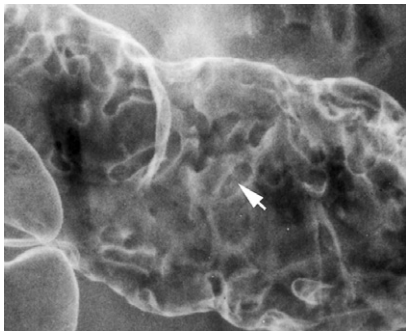
Toxic megacolon. Luminal dilatation, abnormal haustration, mural thickening and mucosal islands. Mucosal islands represent oedematous mucosal remnants.†



Stratified wall thickening in UC on US. The outer low reflective muscle layer is well defined, but the thickened mucosa/submucosa are poorly distinguished. The mucosal surface is indicated by the bright central reflective line (arrow).*



DCBE in an acute attack of UC with collar stud ulcers (arrow) protruding through the mucosal line (arrowhead).*



Filiform postinflammatory polyposis (arrow) following an acute attack of UC. The mucosal surface and haustration are normal as the colitis was inactive.*

Distinguishing features between ulcerative and Crohn's colitis*

Radiographic feature	Ulcerative colitis	Crohn's disease
SB involvement	Reflux ileitis only	+++
Rectal involvement	Always	50%
Multiple anal fistula	–	+
Aphthoid ulceration	–	+++
Fissuring ulceration	–	++
Granularity	+++	+
Transverse symmetry	Symmetric	Asymmetric
Longitudinal extent	In continuity	Discontinuous
Free perforation	+	–
Toxic megacolon	+	–/+
Cancer risk	+	–/+
Entero-enteric fistula	–	+
Submucosal inflammation	++ in chronic disease	–
Mesenteric inflammation	–/+	++
Enlarged lymph nodes	–	+
Fibrofatty proliferation	Mesorectal only	++

ISCHAEMIC COLITIS

Definition This is a common cause of colitis in the elderly, often involving the splenic flexure or proximal descending colon (the 'watershed' areas) ► it is associated with bacterial superinfection

- *Mild disease:* initially the mucosa is the most susceptible to vascular compromise (with oedema, haemorrhage, or necrosis) ► recovery is usually complete
- *Severe disease:* necrosis of the submucosal and muscle layers leads to fibrosis and stricture formation ► transmural necrosis is life-threatening (due to the risk of perforation)

Clinical presentation Disease commonly affects the SMA or IMA distributions:

- *SMA:* these are very unwell patients (they are acidotic ± abdominal pain) ► it often requires surgery
- *IMA:* this has a less acute presentation (and can mimic diverticulitis) ► it can be treated conservatively

AXR A narrowed colon (secondary to stricture formation) ► mucosal thumb-printing (due to submucosal haemorrhage and oedema) ► free air (secondary to perforation) ► colonic sacculation

CT Colonic wall thickening (this is more marked with venous occlusion but does not correspond with the extent of necrosis) ► submucosal oedema can cause a 'target' sign

- *Transmural necrosis:* mesenteric fat stranding ► free fluid ► pneumatosis ► portomesenteric gas
- *'Shock' bowel:* there is increased mural enhancement (due to the generalized low perfusion state) ► there can be a 'slit-like' IVC (as a result of the low intravascular volume)

Pearls Causes of ischaemic colitis:

- *Mesenteric occlusion:* this can be arterial or venous in origin
- *Mechanical obstruction:* secondary to bowel strangulation or obstruction
- *Low flow states*

INFECTIOUS COLITIS

- **Salmonella:** there may be a marked ileus during the acute stage ► a toxic megacolon has been reported
- **Shigella:** this usually affects the sigmoid colon (with aphthoid-type ulceration)
- **Campylobacter:** this affects the distal colon
- **Gonococcus:** this usually affects the rectum
- **Amoebiasis:** this usually affects the right colon and caecum
  - It leads to a segmental or diffuse colitis with granular or ulcerated mucosa (± aphthoid-type ulceration)
  - An amoeboma (an inflammatory granulation mass) is seen in 10% of cases – it can cause irregular stricturing (mimicking a carcinoma)
  - Disease is limited to the caecum in 3% of cases, producing a characteristic conical caecum and a shaggy ulcerated mucosa ► it may be complicated by appendicitis
  - Embolic liver spread is seen in 15% of cases
- **Cytomegalovirus:** this demonstrates an ileocolic distribution ► there is a thick-walled vasculitis with large bleeding ulcers ► mesenteric adenopathy and often ascites is present
- **Herpes simplex virus:** this leads to a proctitis with multiple superficial ulcers
- ***Chlamydia trachomatis:*** this causes lymphogranuloma venereum, which is a chronic proctitis complicated by fistula formation, extensive fibrosis and eventual stricturing

PARASITIC COLITIS

- ***Strongyloides stercoralis:*** this may simulate UC
- **Chagas' disease:** a megacolon results from the neurotoxic effect of the protozoon *Trypanosoma cruzi*
- **Schistosomiasis:** ova are deposited within the large bowel submucosa ► the inflammatory response results in polyp formation ► fibrosis may later cause stricture formation (± bowel wall calcification)

RADIATION COLITIS

Definition This is caused by a radiation-induced occlusive endarteritis with thrombosis and fibrosis ► it is a late complication (often presenting years after radiotherapy when the total dose is > 45Gy)

- *Acute stage:* mucosal injury with an acute colitis
- *Chronic stage:* proctitis (± ulceration) ► strictures are usually smooth and symmetrical unless there is superimposed ulceration ► fistula formation is commonly to the bladder or vagina ► perforation is rare

CT Wall thickening ► increased mesorectal fat and a thickened mesorectal fascia ► a widened presacral space

NEUTROPENIC COLITIS

Definition This is usually due to chemotherapy with bone marrow transplantation

CT A thickened right-sided bowel wall ► pneumatosis ► mesenteric stranding and small bowel involvement is common

- *Typhlitis:* changes limited to the caecum

PSEUDOMEMBRANOUS COLITIS

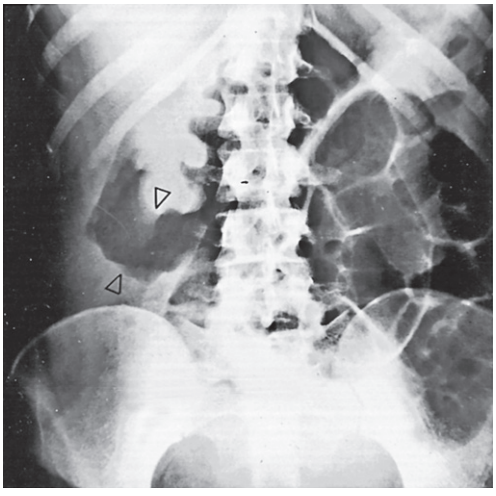
Definition A left-sided or pancolitis due to cytoplasmic endotoxin produced by overgrowth of *Clostridium difficile* ► it is usually as a result of broad-spectrum antibiotic therapy and may be life-threatening

XR A generalized ileus and nodular haustral thickening ► mucosal thumbprinting

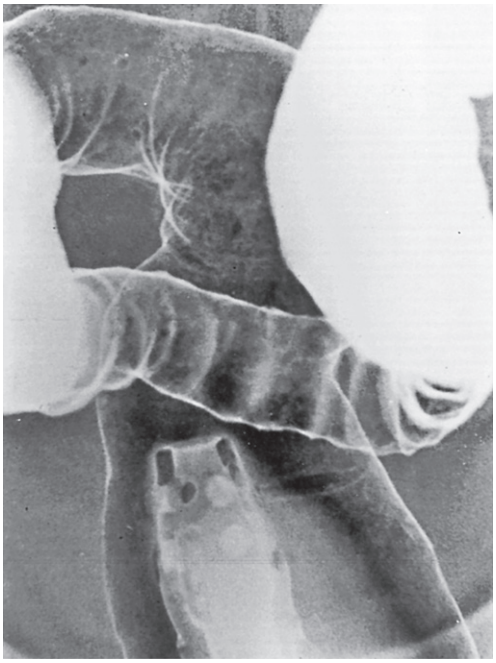
CT Gross colonic wall thickening (average 1.5cm) ► ascites ► small bowel dilatation ► marked mucosal enhancement

- There is minimal pericolonic fat stranding (cf. IBD)
- *'Accordion' sign:* the appearance of contrast between thickened low attenuation mucosal folds

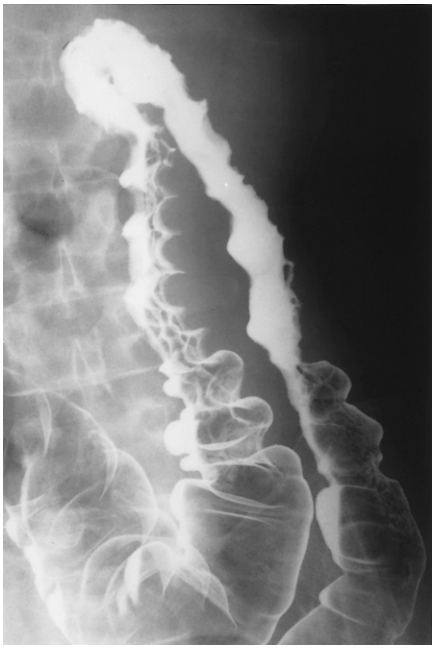
CMV colitis involving the entire colon in addition to the distal small bowel.••



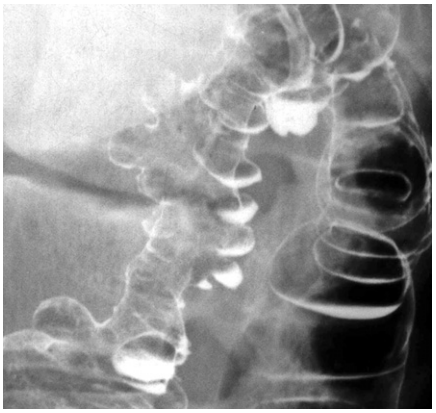
Plain AXR in a patient with fulminant amoebic colitis. Note the narrowed proximal transverse colon and prominent nodularity (thumbprinting) (arrowheads).••



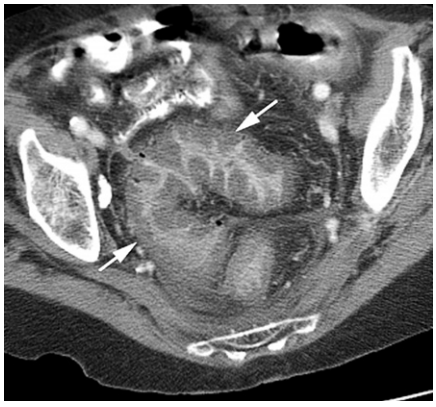
Rectosigmoid spot film in a patient with chronic radiation proctitis and sigmoiditis. There is general loss of haustral pattern with tubular appearance of the bowel.••



Classical splenic flexure 'thumb-printing' due to ischaemic colitis.†



Ischaemic stricture at the splenic flexure with prominent sacculation.*



Pseudomembranous colitis with marked thickening of the colonic wall (arrows) and prominent submucosal oedema creating the accordion sign.*

POLYPS

DEFINITION

- An elevated colonic mucosal lesion

TYPES

Epithelial Adenoma

- This is defined as a circumscribed area of dyplastic epithelium (an intraepithelial neoplasia) ► they are found in 25% of the population over 50 years old (they are rare in patients under 30 years of age)
- An adenoma can be tubular (65%), tubulovillous (25%) or villous (10%) in nature ► villous adenomas may present with electrolyte disturbances due to excessive mucus production
  - *Pedunculated adenoma:* extrusion of a stalk of mucosa and muscularis mucosae may occur as the adenoma is pulled by the faecal stream
  - *Sessile adenoma:* a broad-based lesion (the base must be at least twice that of its height)
  - *Flat adenoma:* a lesion with a height that is no more than twice the height of the adjacent normal mucosa ► it is categorized into a slightly elevated, completely flat, or slightly depressed lesion
- *Location:* rectosigmoid colon (60%) ► descending colon (18%) ► transverse colon (14%) ► ascending colon and caecum (8%)
  - Adenomas tends to be larger when present within the left colon (⅔ of polyps > 2cm are within the rectosigmoid colon)
- Size is the most important single indicator for the likelihood of malignancy:
  - <1cm (<1% risk) ► 1–2cm (10% risk) ► >2cm (approximately a 50% risk)
  - Flat adenomas with a depressed centre may be invasive even if they are small (<1cm)
    - They are more commonly found within the right colon and in association with hereditary non-polyposis colorectal cancer (HNPCC) ► they may demonstrate a more rapid progression to overt cancer than a polypoid tumour

Non-epithelial Carcinoid ► leiomyoma ► lipoma ► fibroma

- *Lipoma:* a well-defined submucosal lesion ► it is usually a solitary right colonic lesion (and may cause pain, bleeding or intussusception if > 4cm)
  - *The 'squeeze' sign:* it is easily deformable during compression
    CT This will easily demonstrate the inherent fat attenuation

Non-neoplastic Juvenile or postinflammatory polyps

Unclassified Hyperplastic polyps – these are usually small with a characteristic 'saw-toothed' epithelial lining ► they are common within the rectum ► there is no malignant potential unless they demonstrate a 'serrated' appearance

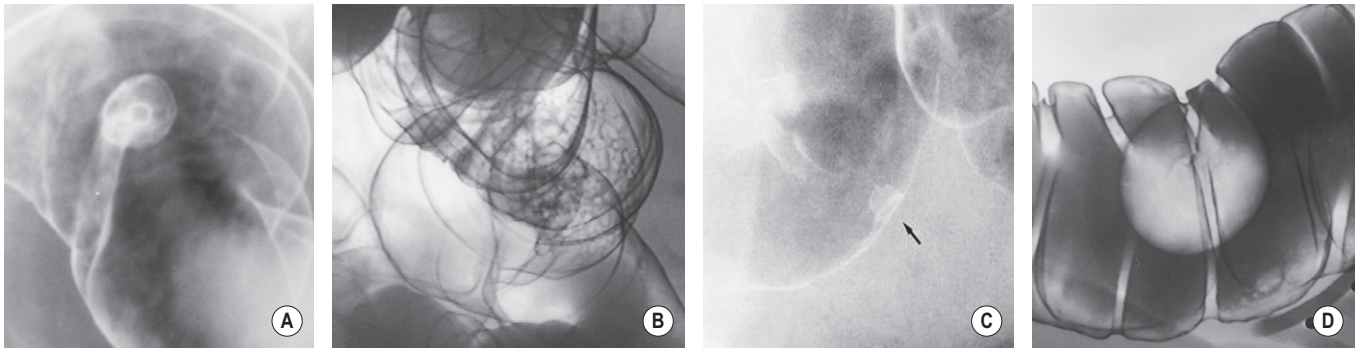
RADIOLOGICAL FEATURES

DCBE appearances of polyps

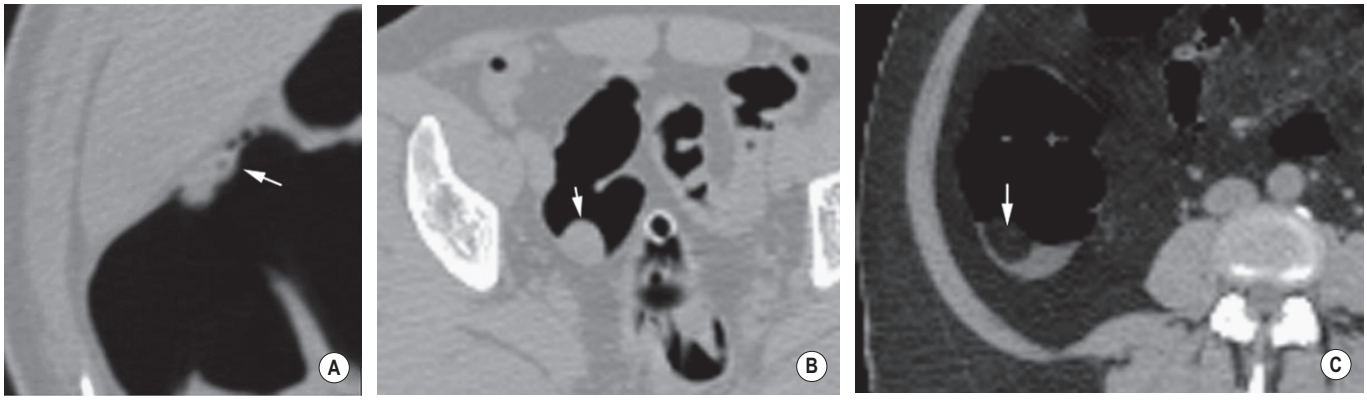
- Localized areas of increased attenuation (the incident X-ray beam passes through more than 2 barium layers and consequently less gas)
- A thin layer of contrast medium covers the mucosa, forming a ring around the polyp base
  - *Viewed en face:* this creates a ring shadow with a sharp inner border and an outer margin fading into the normal surface coating (cf. the opposite appearance with an ulcer)
  - *Viewed obliquely:* this produces the 'hat' sign
- *Pedunculated polyps:* the axis of the stalk usually runs obliquely to the lumen axis (making it easy to distinguish from a haustral fold)
  - *'Stalk' sign:* two parallel lines of barium
  - *'Target' sign:* the head and stalk are superimposed
- *Juvenile polyps:* these are smooth and pedunculated with a thin stalk (affecting patients <40 years old)
- *Postinflammatory polyps:* these have a filiform configuration (i.e. finger-like submucosal projections covered by mucosa on all sides)
- *Villous polyps:* these demonstrate a lace-like or mosaic appearance as barium fills the tumour interstices ► some may present as a flat, nodular, carpet-like growth (with minimal elevation) within the rectosigmoid colon or caecum

CT colonography (CTC) appearances of polyps

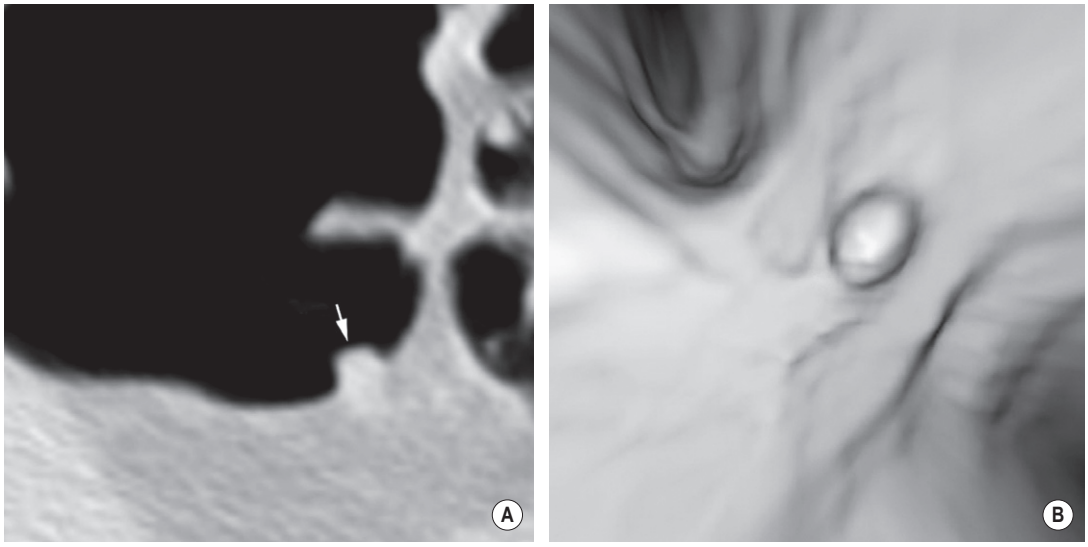
- CTC is equivalent to colonoscopy in the detection of polyps >7mm in size
- Technique: full bowel preparation is standard, although reduced preparation regimens with faecal tagging are being developed ► CO_2 distension is used (and improved with IV Buscopan) ► supine and prone sequences are reviewed to reduce the chance of any collapsed or fluid-filled segments hiding a lesion ► the original datasets and 3D reformatted images (which are useful for problem solving) are reviewed
  - *Pitfall:* an inverted diverticulum may simulate a polyp on 3D images (on 2D images it will contain gas)
- *Distinguishing a polyp from faecal residue:*
  - A polyp demonstrates homogeneous attenuation similar to bowel wall (unless it is a lipoma, when it will be of fat density) ► faecal residue often contains internal gas locules
  - A polyp has a fixed position on the bowel wall – faecal residue will tend to fall onto the dependent colonic surface



(A) When seen en face, stalked polyps produce a 'target' sign. (B) A sigmoid villous adenoma, evidenced by a fine carpeting of frond-like projections. (C) A small polyp where the meniscal rim of barium between the polyp base and adjacent mucosa causes the 'bowler hat' sign (arrow). (D) Transverse colon lipoma. Note its well-defined margins and compressibility under the compression paddle.



CTC appearances. (A) Inhomogeneous attenuation characteristic of faecal residue (arrow). (B) A large sigmoid polyp of homogeneous attenuation (arrow). (C) A lipoma (arrow) demonstrating fat attenuation.



(A) CTC of a small 5mm polyp (arrow) in 2D. (B) CTC of the same polyp in 3D.*

FAMILIAL ADENOMATOUS POLYPOSIS (FAP)

DEFINITION

- An autosomal dominant condition (caused by an APC tumour suppression gene mutation on chromosome 5q21) ► it is characterized by multiple (500–2500) colonic adenomas and requires at least 100 adenomas to be present for the diagnosis to be made
  - The polyps develop by the early teens – all patients will eventually develop colorectal cancer (accounting for 1% of all colorectal cancers) ► a restorative proctocolectomy is recommended once the condition is diagnosed
  - *Associations:* hamartomatous stomach polyps (> 50% of patients) ► duodenal adenoma (almost 100% of patients) ► periampullary carcinoma (5% of patients)

GARDNER'S SYNDROME

- This forms part of the FAP spectrum ► extracolonic manifestations include multiple skull and mandible osteomas, epidermoid cysts, soft tissue tumours, abnormal dentition and desmoid tumours

Desmoid tumour A benign fibromatous tumour involving the abdominal wall or small bowel mesentery ► it is only locally invasive and is often precipitated by surgery

CT Ill-defined mesenteric infiltration with small bowel tethering (giving a 'whorled' appearance) ► this occurs prior to the development of an overt mass ► it can cause ureteric or small bowel obstruction

MRI T2WI: high SI suggests active growth

PEUTZ-JEGHERS SYNDROME

DEFINITION

- An autosomal dominant condition leading to the presence of multiple hamartomas within the stomach, small bowel and colon (colonic polyps are relatively few but are larger, often pedunculated, and may bleed)
  - *Associations:* mucocutaneous pigmentation of the lips, oral mucosa, palms and soles
  - There is no intrinsic malignant potential (although the overlying mucosa may become dysplastic with an increased risk of an upper GI tract cancer)
  - *There is an increased risk of an extraintestinal cancer:* ovary ► thyroid ► testis ► pancreas ► breast

JUVENILE POLYPOSIS

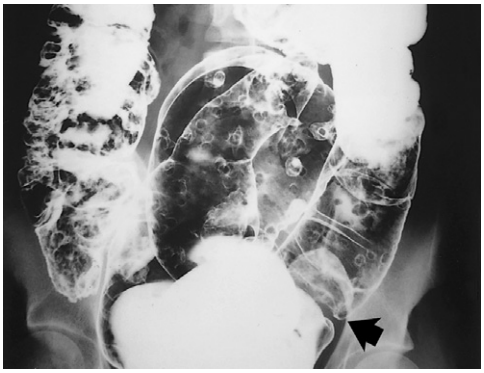
DEFINITION

- Smooth pedunculated hamartomatous polyps are found within the colon (50–200 polyps) as well as the small bowel and stomach ► it is a very rare autosomal dominant condition presenting in infancy
  - *'Swiss cheese' effect:* cystic epithelial tubules in excess of the lamina propria
- Epithelial dysplasia is common in young adults, occurring within either juvenile polyps or co-existing adenomas – there is an increased risk of developing a colorectal carcinoma

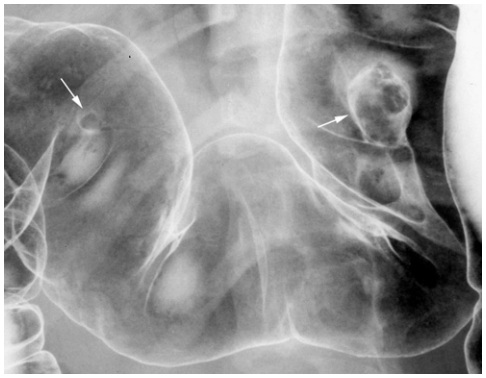
HEREDITARY NON-POLYPOSIS COLORECTAL CANCER (HNPCC)

DEFINITION

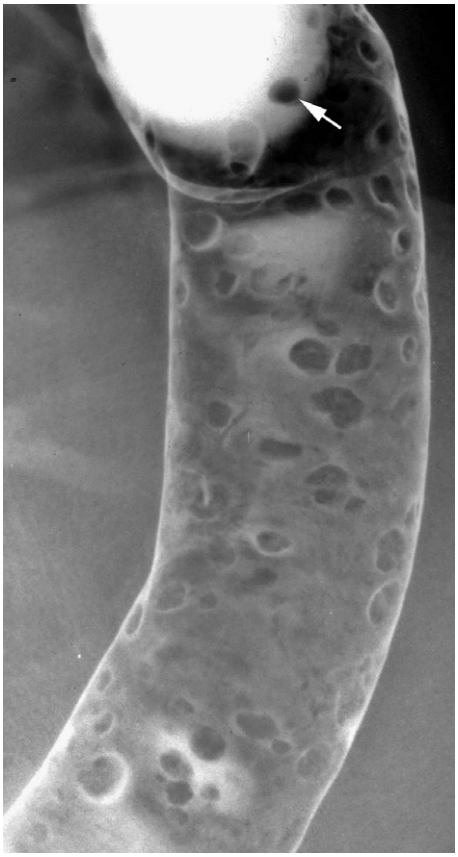
- With HNPCC, polyps are seen from an early age, and cancers occur at an earlier age than those seen in non-HNPCC patients ► the diagnosis requires:
  - $\geq$ 3 relatives with colorectal cancer (one of which is a 1st-degree relative)
  - Cases over 2 or more generations
  - A colorectal carcinoma diagnosed before the age of 50 years
  - *Location:* most lesions are within the proximal colon (70%) ► multiple tumours are common
  - *Associations:* cancers of the breast, endometrium, ovary and pancreas



FAP – there are innumerable colonic adenomas. The patient refused surgery, with the inevitable consequence of a cancer (arrow).†



Peutz–Jeghers syndrome with a large pedunculated and smaller sessile polyp proximally (arrows).*



DCBE view of the descending colon in FAP with multiple small polyps about 5mm in size, creating ring shadow menisci around their bases, or as a filling defect in the barium pool (arrow).*

Other rare polyposis syndromes		
Syndrome	**Inheritance**	**Manifestations**
Turcot's syndrome	Autosomal recessive	An association between colonic carcinoma, polyps and medulloblastoma
Cowden's syndrome	Autosomal dominant	Hamartomatous intestinal polyposis and lesions of the skin, mucous membranes, breast and thyroid
Muir–Torre syndrome	Autosomal dominant	Benign cutaneous sebaceous adenomas and keratoacanthomas ► it is associated with GI polyps and cancers in various sites
Cronkhite–Canada syndrome	Non-hereditary	Diffuse intestinal polyposis (usually colonic although the stomach and small bowel can be affected) ► it is associated with alopecia, skin hyperpigmentation and nail atrophy secondary to malabsorption ► it is rapidly fatal

Classification of polyps and polyposis syndrome*		
Histological type	**Single or few in number**	**Polyposis**
Epithelial	Adenoma – tubular, villous, tubulovillous Adenocarcinoma	Familial adenomatous polyposis, Turcot's syndrome, Cowden's disease
Harmatomatous	Juvenile Metaplastic	Juvenile polyposis Peutz–Jeghers syndrome Metaplastic polyposis
Inflammatory	Postinflammatory polyp	Postinflammatory polyposis
Non-epithelial	Lipoma, carcinoid, GIST, benign lymphoid, neurofibroma	Lymphomatous polyposis, metastatic neurofibromatosis
Miscellaneous	Endometriosis	Cronkhite-Canada syndrome

COLORECTAL CANCER

Definition

- Extension of a malignant intraepithelial neoplasia (adenoma) into the submucosa
- It generally develops from a polypoid adenoma over many years via a multistep accumulation of genetic faults (the adenoma to carcinoma sequence)
  - Familial adenomatous polyposis (FAP) accounts for 1%, and hereditary non-polyposis colorectal cancer (HNPCC) for 5–10% of all cases
  - There is also an increased risk with inflammatory bowel disease, obesity, red meat consumption, smoking and excess alcohol consumption
- Lifetime risk: 1 in 18 (men) ► 1 in 20 (women) ► 65% of cases are >60 years

Clinical presentation

- Change in bowel habit ► rectal bleeding ► abdominal pain ► bowel obstruction (<20% of cases)
  - *Generally:* fresh blood = a distal lesion ► altered blood or anaemia = a proximal lesion (e.g. involving the caecum)

Radiological features

Double-contrast barium enema This only demonstrates the luminal aspect of a tumour (tumour spread is therefore difficult to assess)

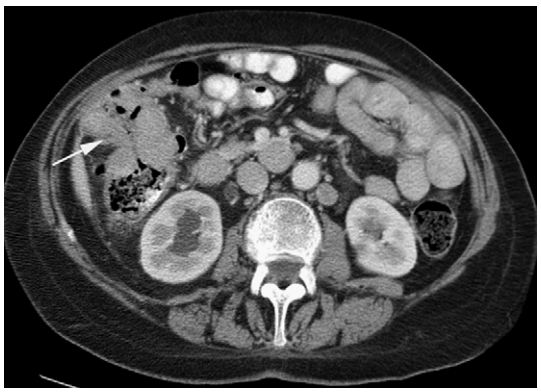
- *Early:* a sessile (plaque-like) lesion, or a pedunculated lesion
- *Late:* polypoid cancers have an irregular in-drawn base ► carpet lesions are seen with malignant villous tumours
  - *'Apple-core' lesion:* an annular or semi-annular lesion with abrupt shouldered margins and an irregular narrow lumen

CT ***This is generally used for assessing the presence of any metastatic disease and not for determining the local T staging***

- It demonstrates luminal and extraluminal disease, the extent of any wall thickening (normal colonic wall is <4mm), and luminal narrowing ► tumour may appear as a focal soft tissue mass (± necrosis)
- *Extramural infiltration*: this is suggested by (but not pathognomonic for): irregular projections from the serosal surface ► clouding of the pericolic fat ► loss of the normal fat planes ► thickened contiguous fascial reflections
- *Tumour enhancement*: this is usually homogeneous ► heterogeneous enhancement can be seen with an abscess, a large adenocarcinoma or a mucinous tumour
- *Intratumour calcification:* this is seen with mucinous adenocarcinomas
- *Enlarged nodes:* these can can be due to reactive hyperplasia or metastatic involvement
  - A metastasis is suggested if a node measures >1cm diameter (short axis) or if there is a cluster of >3 nodes
- *Intraperitoneal spread is indicated by:* ascites ► peritoneal deposits ► omental cake
- *CT colonography:* this can be used to screen asymptomatic patients, for surveillance in high-risk patients, and for evaluating symptomatic patients following a failed colonoscopy
- *Frail elderly patients:* a minimal preparation CT (1.5L 1% Gastrografin 48 h prior to imaging) has an 85% sensitivity for detecting CRC

MRI ***This is used for the local staging of rectal cancer*** (which is considered a separate entity to colon cancer as its pelvic location reduces the ability to obtain wide resection margins with a consequent increased risk of local recurrence)

- *It can assess for:* extramural spread ► peritoneal infiltration ► extramural venous involvement ► nodal involvement ► response to chemoradiotherapy
- **Bowel wall layers (T2WI):** *muscularis mucosa:* a fine low SI line ► *submucosa:* a thicker high SI layer ► *muscularis propria:* inner circular and outer longitudinal layer with an irregular grooved appearance ► *perirectal fat*: high SI ► *mesorectal fascia:* a fine low SI layer enveloping the perirectal fat
- **Mesorectal fascia:** this encloses the mesorectum (which contains the draining lymphatic nodes and vessels) ► nodal spread usually occurs cranially within this compartment ► caudal spread and pelvic side wall involvement is unusual (caudal spread and associated inguinal lymph node involvement can occur if there is some impairment of the normal drainage pattern) ► a total mesorectal excision (TME) without breach of the mesorectal fascia minimizes local recurrence
  - *Positive circumferential resection margin (CRM):* if there is tumour within 1mm of the mesorectal fascia this requires preoperative chemotherapy
- **Local nodal involvement:** small nodes may still be involved by tumour ► malignant nodes tend to have an irregular border and demonstrate internal mixed SI ► it is important to record whether a suspicious node lies within 1mm of the CRM
- **Extramural vascular invasion:** this describes the presence of tumour cells beyond the muscularis propria and within endothelial-lined vessels ► any extramural vascular invasion is further classified according to the number of vessels involved and whether they can be identified anatomically
- **Poor prognostic features:** increasing depth of extramural invasion ► nodal involvement ► involvement of the circumferential resection margin ► extramural vascular invasion

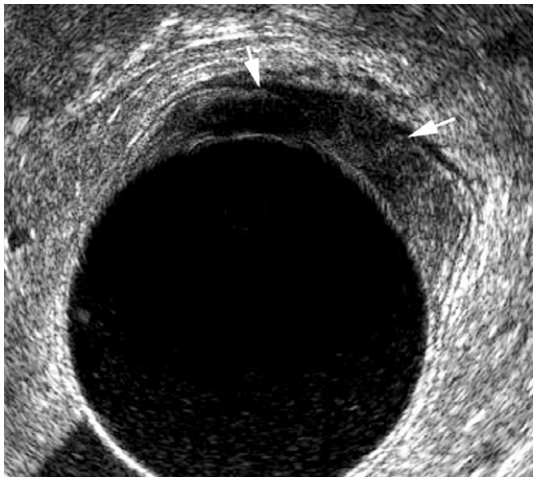


Unprepared CT showing a T3 carcinoma (arrow) in the hepatic flexure with fascial thickening. Note also the enlarged para-aortic nodes.*

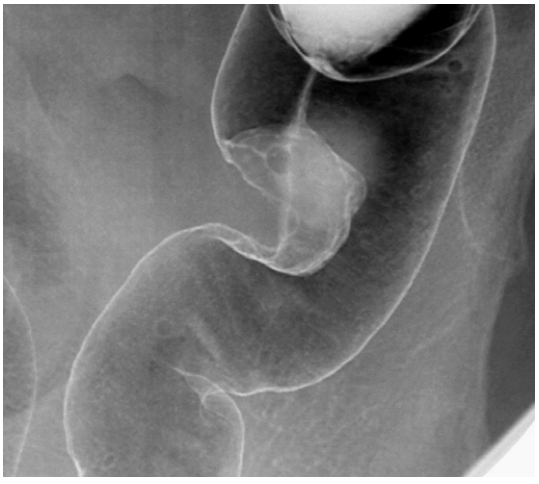
Typical 'apple-core' sigmoid carcinoma.†

Axial T2WI of the pelvis with a cancer extending outside the rectal wall (white arrow), a large heterogeneous nodal mass posteriorly touching the posterior mesorectal fascia (arrowhead), and an involved mesorectal node to the left of this (T3 N1 stage).*

Axial T2WI of the pelvis showing the mesorectal fascia (arrows).



Endorectal US of a T2 rectal cancer. The submucosa is breached (white arrows) but the muscularis propria is intact.



A polypoid carcinoma with an irregular in-drawn base.*

COLORECTAL CANCER

FDG PET-CT

- This is useful for detecting extracolonic disease (lesions >1cm) ► it is the most accurate modality for detecting recurrent pelvic cancer (scar tissue does not demonstrate increased FDG PET uptake)

Endorectal US

- This allows for better differentiation of the rectal wall layers than MRI ► it is used to assess early tumour involvement and select T1N0 cases for local excision

PEARLS

Approximate distribution Rectum (35%) ► sigmoid colon (25%) ► descending colon (10%) ► transverse colon (10%) ► ascending colon (10%) ► caecum (10%)

- Right-sided tumours are more common in the elderly
- A primary scirrhous carcinoma with a pronounced desmoplastic reaction is very rare (and is indicated by long circumferential spread)

Liver metastases

- *Colon and upper rectum:* drainage is via the portal vein and therefore commonly leads to liver metastases (however metastatic deposits derive their blood supply from the hepatic artery and will therefore appear hypoattenuating on portal phase imaging)
- *Lower rectum:* drainage is via the portal vein as well as directly into the IVC (via the pelvic veins) ► metastases can therefore be to the liver but can also produce isolated pulmonary metastases (without liver involvement)
- *Mucinous tumours:* these produce cystic or calcified liver metastases ► there can be widespread peritoneal deposits
- *Liver-specific MR contrast agents:* Mn-DPDP is taken up by functioning hepatocytes, leading to a prolonged T1WI SI
  - T1WI: the conspicuity of any metastases (demonstrating relatively reduced SI) is increased
- A partial hepatic resection can remove all anatomically resectable liver metastases as long as:
  - sufficient normal liver tissue remains to allow normal hepatic function
  - there is no extrahepatic disease
- *Other common metastatic sites:* adrenal glands ► bone (lytic deposits)

Treatment Surgical resection for localized disease ± neoadjuvant therapy ± adjuvant therapy (depending upon the recurrence risk) ► chemotherapy or radiotherapy for non-resectable disease

Emergency radiologically guided colonic stenting For acute obstruction or palliation

Differential Diverticulitis (which can appear very similar to a colonic tumour, especially if a tumour has perforated) ► ischaemic colitis ► inflammatory bowel disease ► local colonic spasm

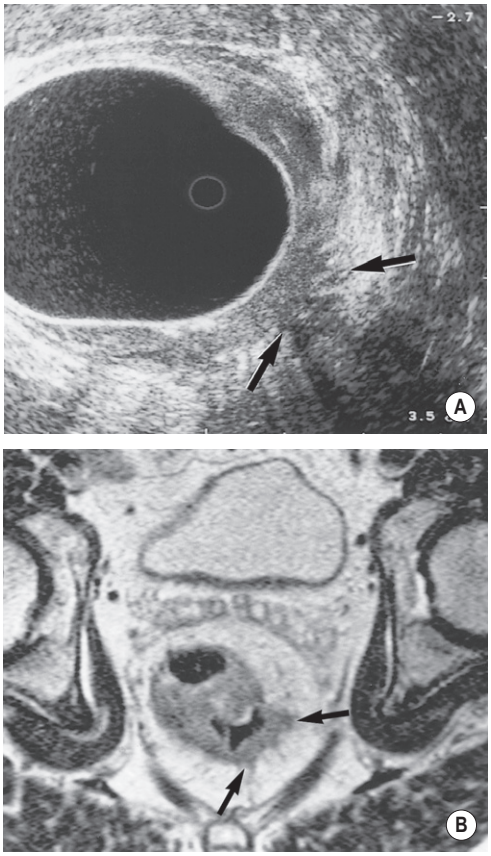
Complications Colonic obstruction (with advanced disease) ► tumour perforation (with an adverse prognostic effect due to tumour dissemination and any local peritonitis) ► colonic intussusception (the tumour acts as the lead point) ► appendicitis presenting in the elderly (due to appendiceal obstruction by a caecal tumour)

Disease recurrence This commonly presents with pelvic pain ► rising CEA levels should prompt a search for recurrence ► it leads to a poor prognosis as most cases are unresectable

- *Common sites:* at the anastomotic site ► within the presacral region with soft tissue thickening (post-surgical changes can cause significant scar formation – therefore an early postoperative scan is essential to act as a baseline) ► a growing mesorectal nodule ► nodular thickening of the pelvic side wall ► peritoneal seeding (which is commonly seen within the pouch of Douglas or the lower right small bowel mesentery)
- **MRI:** T2WI: recurrence demonstrates higher SI than fibrosis (image after the first 6 months to allow for post-surgical resolution) ► there is possibly increased enhancement (and washout) demonstrated by tumour recurrence vs fibrosis
- **FDG-PET:** recurrence demonstrates increased uptake (fibrosis does not)

Dissemination of other tumours to the colon via

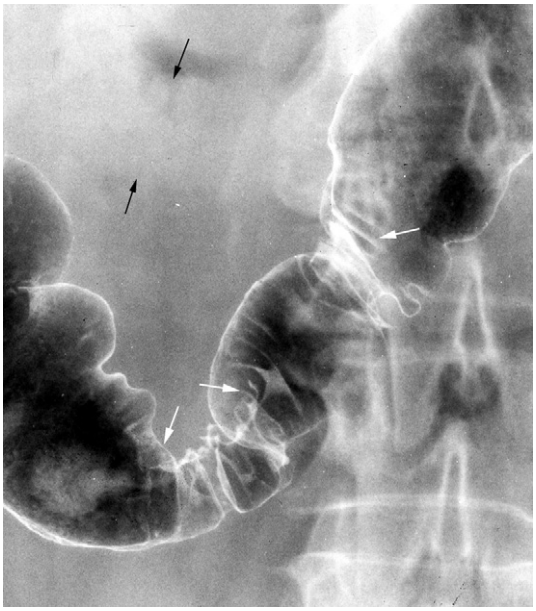
- *Direct invasion:* a gastric carcinoma may invade the transverse colon via the gastrocolic ligament and a pancreatic cancer via the transverse mesocolon (with superior and inferior transverse colonic border changes, respectively)
  - *Serosal involvement:* there is mass effect with a spiculated fold pattern due to the associated desmoplastic response
- *Haematogenous spread:* breast carcinoma and melanoma tumour cells have a propensity to spread to the colon ► tumour cells embolize to the colonic vasa recta capillaries (presenting as submucosal masses on the antimesenteric border) ► these can be polypoid or umbilicated (due to a differential growth pattern between the centre and the periphery)
- *Along the mesenteric planes ► via lymphatic spread ► via intraperitoneal seeding*



(A) Transrectal US reveals a T3 posterior tumour that has penetrated the muscularis propria to reach the surrounding tissue (arrows). (B) T2WI confirms the rectal wall penetration (arrows).

CTC of an annular carcinoma revealing the irregular lumen and thickened bowel wall (arrows).

MRI T staging of rectal cancer	
T1	Intermediate SI involving mucosa/submucosa
T2	Intermediate SI within the muscularis propria
T3	Intermediate SI nodularity projecting beyond the muscularis propria and into the perirectal fat
T4	Abnormal SI projecting beyond the serosa into an adjacent organ or structure, or tumours that have perforated the peritoneum



Transverse colonic serosal deposits causing spiculation and mass effects on the superior border (white arrows) due to local spread from a gastric cancer (black arrows) via the gastrocolic ligament.

Self-expanding metal stent crossing a low rectal tumour.

Duke's classification	Description	5-year survival
A	Tumour limited to the rectal wall	85–95%
B	Tumour spread to the extrarectal tissues, no nodal spread	60–80%
C	Nodal spread	30–60%

DIVERTICULOSIS

Definition

Diverticulum A pouch of mucous membrane with a very thin covering of longitudinal muscle ► it arises between the mesenteric and antimesenteric taeniae at the points of weakness where the vasa recta penetrate the circular muscle layer

Diverticulosis The presence of colonic diverticula

Diverticulitis Inflammatory changes within one or more colonic diverticula (10% of patients with diverticulosis)

- It is caused by faecal retention within a diverticulum – this leads to ischaemic necrosis (± perforation)
- *Complications:* faecal peritonitis (rare) ► colonic bleeding due to weakening of the vasa recta walls – this can be profuse ► abscess and fistula formation

Location Diverticulosis is commonest within the sigmoid colon ► localized to the proximal colon in only 10% (right-sided diverticulae tend to be larger with wider mouths)

- It is extremely rare within the rectum

Radiological features

CT Increasingly used as the initial investigation:

- *Diverticulosis:* multiple outpouchings from the bowel wall (± bowel wall thickening)
- *Diverticulitis:* diverticulosis + inflammation: mural thickening ► pericolic 'fat stranding' and oedema producing a generalized increase in attenuation ► colonic obstruction (10% perforation)
  - *Mild disease:* minimal wall thickening (4–5mm) ► inflammatory changes within pericolic fat only
  - *Moderate disease:* abscess formation
  - *Severe disease:* wall thickening >5mm ► perforation ► large abscess (>5cm) formation ► inflammatory extension into the pelvis
- *Pericolic abscess* (35%): a localized fluid collection with enhancing walls – communication with the bowel lumen is confirmed by gas within the abscess (± extravasation of any luminal contrast)
- *Fistula formation:* this is usually with the bladder (with focal bladder wall thickening)

Pearls

- It can be difficult to distinguish diverticulitis from a colonic cancer: diverticulitis will tend to demonstrate pericolic inflammation and colonic involvement >10cm ► cancer will tend to demonstrate enlarged nodes and a discrete mass
- **Giant sigmoid diverticulum:** a large gas-filled structure that is rarely seen within the lower abdomen
  - *Complications:* diverticulitis ► small bowel obstruction ► perforation ► volvulus
- **Epiploic appendagitis:** this follows infarction of an epiploic appendage, causing acute pain similar to that seen with appendicitis or diverticulitis ► it usually resolves spontaneously
  - **US:** a non-compressible pericolic hyperechoic ovoid mass immediately under the abdominal wall
  - **CT:** a focal area of hyper-attenuation with a central area of fat density

LARGE BOWEL STRICTURES

Definition

- Colonic strictures (narrowing) can be due to various causes (see Table):
  - It is important to distinguish a stricture from a functional narrowing – 7 'physiological sphincters' exist within the colon (e.g. Cannon's point within the mid-transverse colon)

Radiological features

Double-contrast barium enema

- *Fibrotic strictures:* classically this has a smooth lumen with tapering ends
- *Malignant strictures:* classically this has an 'apple-core' configuration: an irregular lumen with shouldered ends involving a short colonic segment
- *Diverticular disease:* narrowing is common ► it is distinguished from a malignant stricture by retention of its mucosal folds and demonstration of the spiculated necks of compressed diverticulae

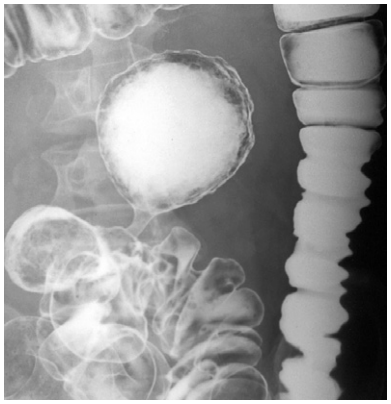
Further clues to the underlying aetiology (on DCBE and CT)

- *Wall thickening:* minimal and symmetric (with ischaemia) ► marked (with an inflammatory mass) ► eccentric (with tumour)
- *Sacculation:* only with ischaemia or Crohn's disease
- *Contraction of the mucosal folds:* crenation with a mass effect is characteristic of a desmoplastic response with endometriosis or carcinoma
- *Site of the stricture:* rectosigmoid (radiotherapy) ► anterior rectosigmoid wall (endometriosis) ► splenic flexure (ischaemia) ► disease elsewhere (Crohn's disease)
- *Extraluminal components:* associated fibrosis (radiotherapy) ► lack of a discrete extraluminal component (ischaemia)
- *Malignant potential:* mesenteric stranding suggests a likely benign cause ► nodal enlargement suggests a likely malignant cause

Acute diverticulitis with thickening of the wall of the sigmoid and marked inflammatory changes in the surrounding mesenteric fat. Gas is seen in several diverticula.*

CT of diverticulitis with a large pelvic abscess containing an air-fluid level (arrows) indicating a communicating abscess. Note the diverticulum outlined by air (arrowhead).*

CT of a bladder fistula secondary to diverticulitis. Note the gas in the bladder indicating a fistulous communication to the bowel, the presence of sigmoid diverticular disease with inflammatory thickening of the base of the bladder at the site of the fistula.*



A giant sigmoid diverticulum with sigmoid diverticular disease.*

Splenic flexure sacculation and stricturing as sequelae to ischaemic colitis.†

Causes of large bowel strictures	
Physiological	Distended bladder ► spasm
Surgical	Anastomosis ► site of colostomy
Malignant	Annular ► scirrhous ► metastatic carcinoma ► lymphoma
Diverticular disease	Pericolic abscess
Ischaemia	Sacculation common as with Crohn's strictures
Radiation colitis	In radiation field so usually rectosigmoid
Inflammatory bowel disease	Ulcerative colitis ► Crohn's disease ► tuberculosis ► lymphogranuloma venereum ► amoebiasis
Miscellaneous	Extrinsic masses ► endometriosis ► pelvic lipomatosis ► trauma

MECHANICAL LARGE BOWEL OBSTRUCTION (LBO)

Definition

- There are numerous causes: colonic carcinoma (the commonest cause) ► diverticulitis (the 2nd commonest cause) ► colonic volvulus
  - Obstruction more commonly involves the left colon
  - Adhesive large bowel obstruction is very unusual (cf. small bowel obstruction)

Radiological features

AXR/instant unprepared barium enema/CT Appearances depend on the site of the obstruction and whether the ileocaecal valve remains competent:

- *A competent ileocaecal valve:* this affects a minority of patients ► in spite of increasing intracolonic pressure and marked caecal distension, the small bowel is not distended
- *An incompetent ileocaecal valve:* there is marked small bowel dilatation ► the caecum and ascending colon are not unduly distended

Pearls

- If both small and large bowel dilatation is present then the appearances can mimic a paralytic ileus
- As large bowel obstruction can mimic a pseudo-obstruction, any patient with a suspected large bowel obstruction therefore requires further imaging (e.g. an instant unprepared enema) to confirm the diagnosis
- There is a risk of perforation if the caecum measures >9cm (and the transverse colon >6cm)

LARGE BOWEL VOLVULUS

Definition

- Twisting of a colonic segment around its mesenteric attachment – it can therefore only occur in those parts of the colon that have a long freely mobile mesentery (sigmoid colon > caecum > transverse colon)
- **Caecal volvulus:** this can only occur when the caecum and ascending colon are on a mesentery (this is not seen in all patients) ► it affects a younger age group (30–60 years) than a sigmoid volvulus
- **Sigmoid volvulus:** the sigmoid colon twists around its mesenteric axis ► it is usually chronic with intermittent acute attacks ► it tends to occur in old age or in patients with a mental handicap or institutionalization

Radiological features

Caecal volvulus

AXR/CT A gas-filled distended caecum ► the haustra are still visible (cf. sigmoid volvulus) ► often there is small bowel distension (the left side of the colon is usually collapsed)

- The caecum can twist and invert with the caecal pole and appendix occupying the left upper quadrant
- The caecum can twist in the axial plane without inversion – the caecum then occupies the right lower quadrant ► this is associated with vascular compromise

Sigmoid volvulus

AXR/CT A massively air distended inverted U-shaped loop ► proximal colonic dilatation is typical ► the apex of the loop usually lies above T10 and under the left hemidiaphragm ► the margins of the loops are devoid of haustra ► there is generally an air:fluid ratio of >2:1

- *'Inferior convergence':* the two limbs of the loop converge inferiorly on the left at the level of the upper sacral segments
- *'Liver overlap' sign:* the ahaustral margin overlaps the lower liver border
- *'Left flank overlap' sign:* the ahaustal margin overlaps the haustrated dilated descending colon
- *'Pelvic overlap' sign:* the ahaustral margin overlaps the left side of the pelvis

Contrast enema/CT The mucosal folds can show a 'screw' pattern at the point of twisting

- *'Bird of prey' sign:* the point of torsion appears as a smooth, curved tapering of the colonic lumen, which can look like a hooked beak

PARALYTIC ILEUS

Definition

- Peristalsis ceases with accumulation of fluid and gas within the bowel

Radiological features

AXR The appearances can vary from dilatation of a short length of small bowel (e.g. following localized pancreatitis) to dilatation of the entire intestine (e.g. following peritonitis) ► it can be difficult to differentiate from a low large bowel obstruction

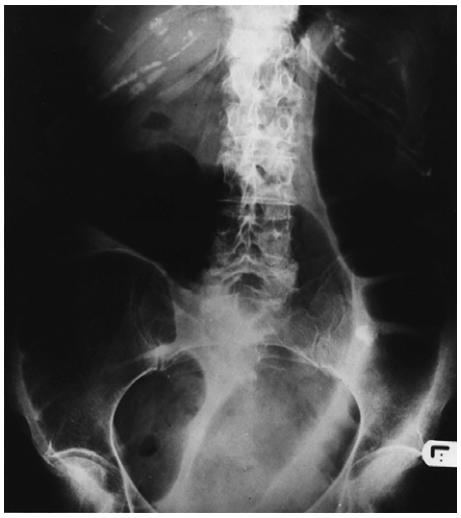
PSEUDO-OBSTRUCTION (OGILVIE'S SYNDROME)

Definition

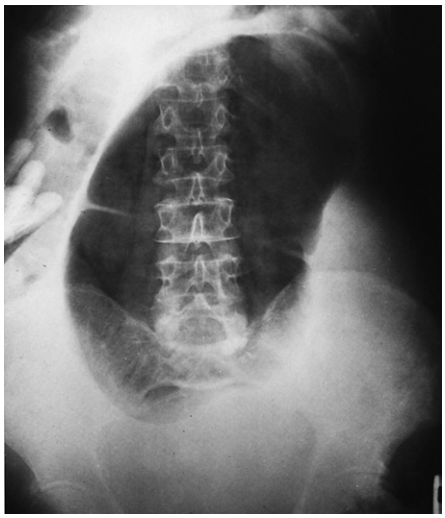
- This usually occurs in elderly patients and is often due to cathartic abuse ► there is no mechanical obstruction but it will mimic intestinal obstruction clinically and radiologically
- Mechanical obstruction needs to be excluded with a contrast enema, CT or colonoscopy

Pearls

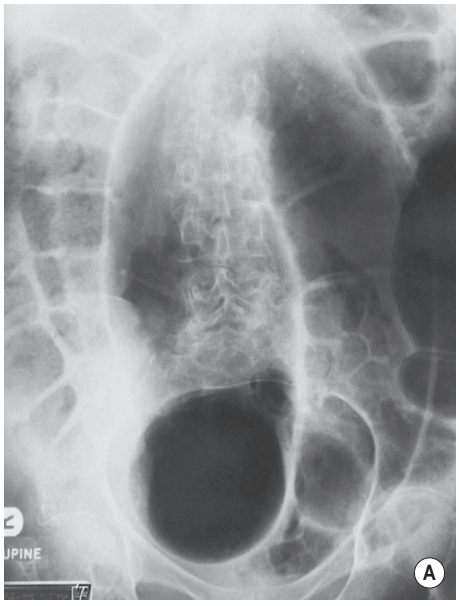
- The caecum may exceed the critical diameter of 9cm with the risk of imminent perforation – a caecostomy or right-sided colostomy may be urgently required

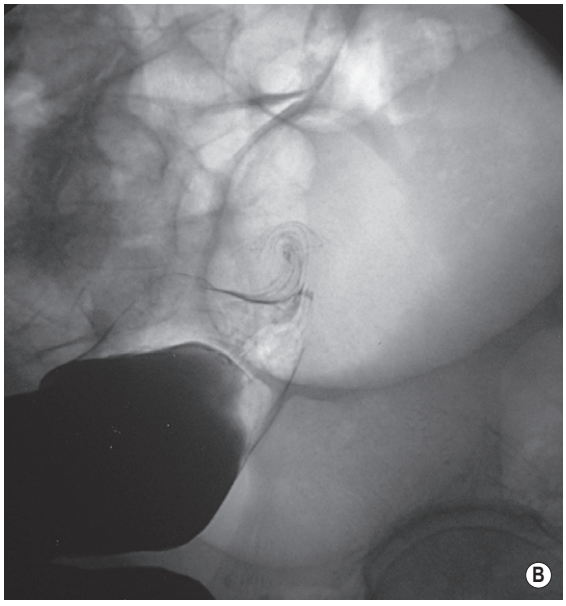


Large bowel obstruction in sigmoid carcinoma: supine position. Gas-filled, distended large bowel and caecum. A competent ileocaecal valve has resulted in no dilatation of small bowel.*

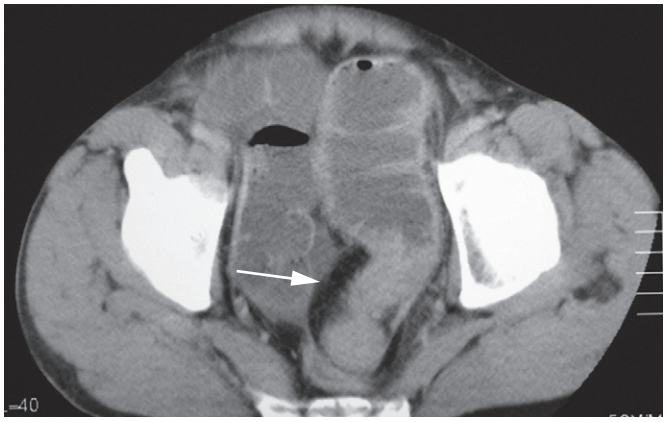


Caecal volvulus. The dilated caecum lies with its pole under the left hemidiaphragm. In spite of the dilatation the haustra are preserved. There is no dilated large bowel elsewhere in the abdomen. The small bowel is fluid filled in this case.*





Sigmoid volvulus. (A) Massively dilated distended gas-filled loop of sigmoid colon. (B) Contrast enema showing the twisted sigmoid colon (bird of prey sign).*



Large bowel obstruction. CT demonstrating dilated fluid-filled large bowel due to a stricture of the sigmoid colon (arrow). Histology of resection specimen showed stricture due to diverticulitis.*

Causes of a paralytic ileus	
Peritonitis	Congestive heart failure
Surgery	Pneumonia
Trauma	Renal colic
- *spine*	Renal failure
- *ribs*	Leaking abdominal aortic aneurysm
- *hip*	Low serum potassium
- *retroperitoneum*	Drugs (e.g. morphine)
Inflammation	Spinal lesions
- *appendicitis*	General debility or infection
- *pancreatitis*	Vascular occlusion
- *cholecystitis*	
- *salpingitis*	

ACUTE APPENDICITIS

Definition

- Inflammation of the appendix following obstruction of the appendix lumen by an appendicolith, hypertrophied lymphatic tissue or tumour
  - Venous obstruction causes ischaemia with necrosis and bacterial invasion

Radiological features

AXR A localized paralytic ileus ► an associated abscess may lead to indentation of the medial border of the caecum (± loss of the lower part of the properitoneal fat line and right psoas muscle shadow) ► the small bowel may become stuck to the inflamed appendix leading to small bowel obstruction

- An appendicolith can be seen in up to 10% of cases

US The appendix appears as a blind-ending non-compressible tubular structure (with a diameter ≥7mm) ► there is maximal tenderness over the appendix ► an appendicolith will appear as a hyperechoic focus casting an acoustic shadow ► there may also be a hyperechoic inflammatory mass, abscess or free fluid around the appendix

- *A false-negative examination:* focal appendicitis of the appendiceal tip ► a gangrenous or perforated appendicitis
  - ⅔ of appendices have a retrocaecal location (and are therefore difficult to see with US)
- *A false-positive examination:* a dilated Fallopian tube ► peri-appendicitis ► inflammatory bowel disease ► inspissated stool

CT The appendix measures >6mm in diameter ► the appendix will fail to fill with oral contrast medium or air up to its tip ► an appendicolith will appear as a calcified 'stone' ► there may be an enhancing appendiceal wall, local adenopathy, surrounding inflammatory change, an abscess, or extraluminal gas (indicating perforation)

- An appendicolith can be seen in up to 30% of cases
- *'Arrowhead sign':* luminal contrast or air within the caecum and pointing towards the obstructed appendix origin
- *A caecal bar:* focal caecal thickening due to oedema at the appendix origin

Pearl

- **Appendix mucocele:** accumulation of mucus within an appendix due to an aseptic obstruction
  - This results in cyst formation (± mural calcification) ► the cyst may rupture, resulting in pseudomyxoma peritonei

ANAL FISTULA

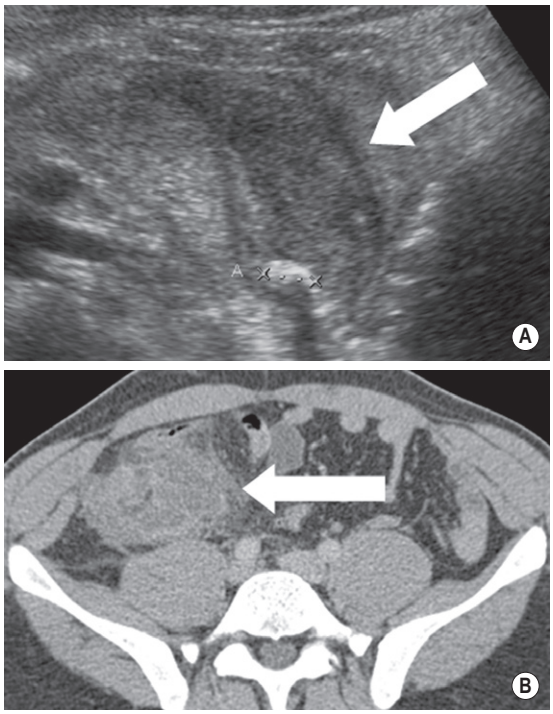
Definition

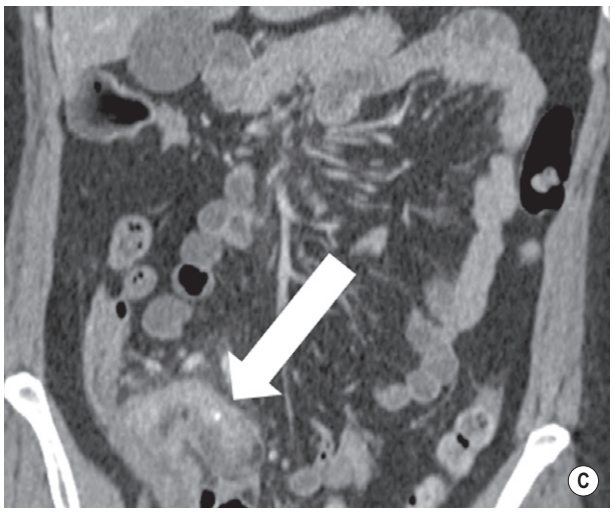
- This is secondary to Crohn's disease or cryptogenic anal gland infection:
  - Discharge of an abscess creates a track through part of the sphincter (usually the longitudinal layer) to the perianal skin ► the internal opening is usually situated posteriorly (at 6 o'clock) and at the level of the dentate line
- **Park's classification:** 'SITES': **S**uperficial ► **I**ntersphincteric ► **T**rans-sphincteric ► **E**xtrasphincteric ► **S**uprasphincteric
- **Goodsall's rule** (on axial imaging):
  - A fistula with an external opening *posterior* to a plane passing horizontally through the centre of the anus: a curved track with its internal opening within the dorsal midline
  - A fistula with its external opening *anterior* to a plane passing horizontally through the centre of the anus: a linear track directly to the nearest anal crypt
  - An external opening adjacent to the anal margin suggests an intersphincteric tract whilst a more laterally located opening suggests a trans-sphincteric tract
  - Openings seen on both sides of the anal canal are likely to arise from a midline posterior crypt with a horseshoe type of fistula
- **Superficial perianal fistula:** the superficial fistula tracks below both the internal anal sphincter and the external anal sphincter complex
- **Intersphincteric fistula (70%):** the intersphincteric fistula tracks between the internal anal sphincter and the external anal sphincter complex in the inter-sphincteric space ► the external opening is at the natal cleft/perianal skin ► the internal opening is usually in the midline posteriorly in the anal canal at the level of the dentate line
- **Trans-sphincteric fistula (25%):** a trans-sphincteric fistula tracks through the internal anal sphincter, inter-sphincteric space and external anal sphincter complex ► the external opening is through the ischioanal fossae to the perianal skin ► the internal opening is into the anal canal at the level of the dentate line
- **Extrasphincteric fistula (1%):** an extrasphincteric fistula tracks outside both internal and external anal sphincters ► it penetrates the levator ani muscle with an internal opening at the level of the rectum
- **Suprasphincteric fistula (5%):** a suprasphincteric fistula travels upwards in the intersphincteric space over the top of puborectalis and penetrates levator ani muscle – it then tracks down to the perianal skin ► the internal opening is into the anal canal at the level of the dentate line

Radiological features

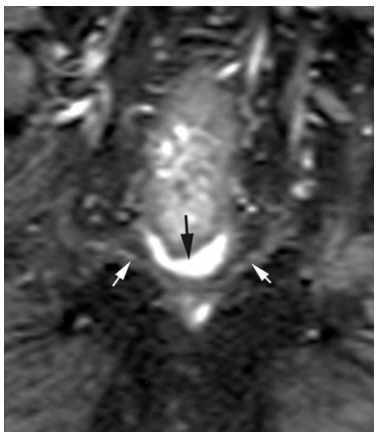
MRI (axial, coronal, sagittal STIR) ► this can demonstrate

- The primary and secondary tracks (high SI on STIR images)
- The internal and external openings
- Any supralevator extension or associated abscess

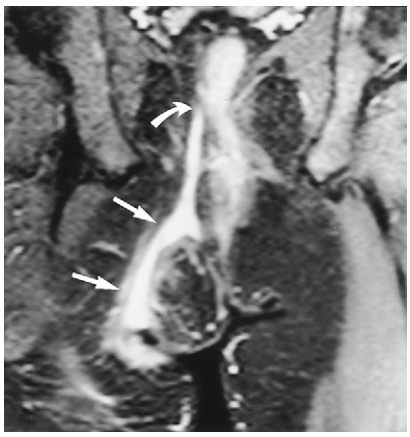




Appendicitis. (A) US demonstrating a thickened appendix wall (arrow), with an appendicolith at its tip. (B) CT demonstrating an appendix mass (arrow). (C) CT demonstrating an inflamed appendix, with a tiny appendicolith as its cause (arrow).

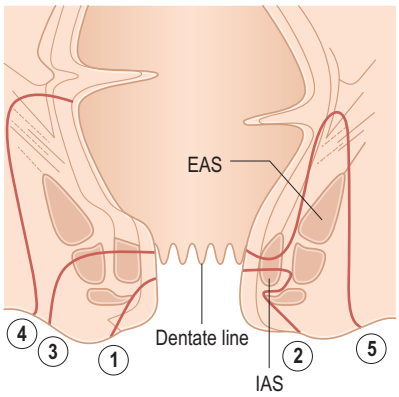


STIR coronal MRI of a supralevator horseshoe abscess (black arrow) above the levator ani (white arrows).*



Coronal STIR MRI reveals a right-sided extrasphincteric fistula (straight arrows) with its enteric communication in the rectum (curved arrow).†

Coronal STIR image of a trans-sphincteric fistula (arrow).*



A classification of fistula-in-ano. EAS: external anal sphincter. IAS: internal anal sphincter.
(1) Superficial
(2) Intersphincteric
(3) Transsphincteric
(4) Extrasphincteric
(5) Suprasphincteric.

CHRONIC MEGACOLON

Definition

- A long-standing dilated colon ► there are various causes:
  - Hirschspung's disease ► chronic laxative abuse ► colonic pseudo-obstruction (Ogilvie's syndrome) ► Chagas' disease ► hypothyroidism ► an electrolyte imbalance ► diabetes ► scleroderma ► amyloidosis

PELVIC LIPOMATOSIS

Definition

- A rare condition of unknown aetiology leading to the proliferation of pelvic adipose tissue

Radiological features

XR Increased radiolucency of the pelvis with exceptionally good sacral delineation

CT/MRI A diffuse increase in the pelvic fat with associated bladder and rectal compression

SOLITARY RECTAL ULCER SYNDROME

Definition

- This forms part of the spectrum of rectal prolapse – it is an area of reddening or ulceration on the anterior rectal wall with associated lamina propria fibrosis ► it presents with difficulty in evacuation, rectal bleeding, and excess mucus production

Radiological features

DCBE Bowel wall deformity at the ulcer site ► mucosal irregularity or nodularity due to the associated granulation tissue

- *Colitis cystica profunda:* polypoid change (due to retention cysts) that may be seen at the ulcer margin

Evacuation proctography Frequently this will show an intra-anal intussusception (which is the cause of the anal trauma)

ENDOMETRIOSIS

Definition

- Gastrointestinal involvement is present in up to ⅓ of cases ► it mainly involves the sigmoid colon, caecum or small bowel loops within the pelvis
  - Serosal implants invade the muscularis propria leading to fibrosis with contraction of the wall and an associated mass effect (the mucosa remains intact)

Radiological features

DCBE A localized mass effect with characteristic contracted mucosal folds

MRI T1WI (FS): high SI ► T2WI: high SI with 'shading' due to the presence of residual blood products (if there is a cystic component)

Differential diagnosis

- Metastatic disease ► chronic pelvic inflammation

LOCAL COMPLICATIONS IN THE POSTOPERATIVE COLON

Definition

- Local complications following colonic surgery: anastomotic breakdown or stricture formation ► postoperative abscess or haematoma formation ► recurrent tumour formation

Radiological features

Water-soluble contrast enema This is performed at about the 12th postoperative day ► it can demonstrate a leak of contrast at the anastomosis or the formation of a stricture

- *Benign stricture*: a smooth outline
- *Malignant stricture:* an irregular outline

CT This can also demonstrate a leak with water-soluble colonic contrast

- *Postoperative abscess:* a rim enhancing fluid collection (± internal locules of air)
- *Haematoma:* a high attenuation fluid collection

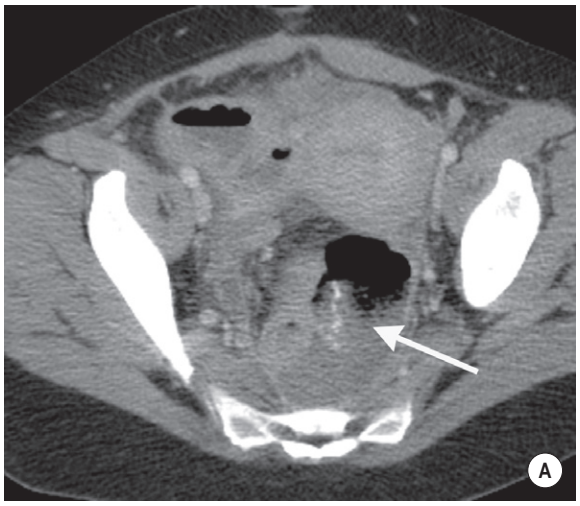
Pearls

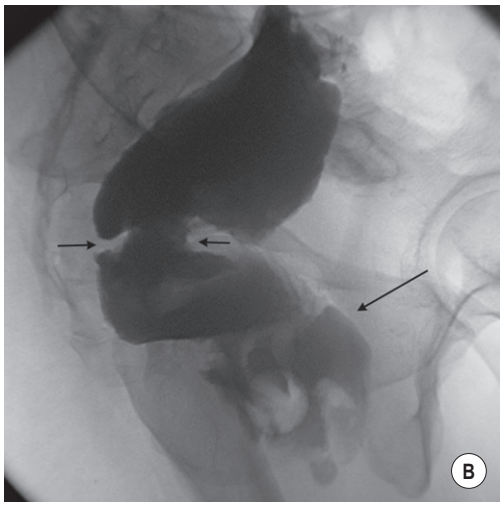
- Colonic resection with anastomosis formation is covered by a defunctioning colostomy – this does not reduce the incidence of a leak but does mitigate against the effects of any associated abscess ► an anastomotic leak warrants delay in closing the colostomy
- A defunctioned colon always has a low-grade bacterial colitis (causing narrowing and loss of haustration)
- Stricture formation is a long-term consequence of anastomotic breakdown
- Anastomotic leakage is a poor prognostic factor for long-term function

RETRORECTAL LESIONS

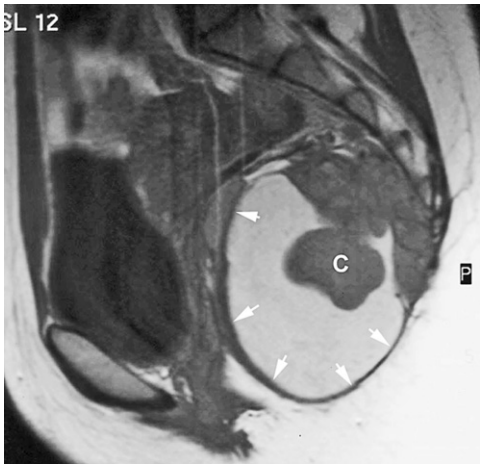
Definition

- These present as a mass that may be complicated by infection, bleeding, or malignant change
  - *Developmental cysts*: epidermoid cyst ► dermoid cyst ► enteric cyst
  - *Sacral lesions*: teratoma ► anterior sacral meningocele ► chordoma ► lymphangioma
  - *Anorectal lesions:* lipoma ► GIST ► anal gland cyst

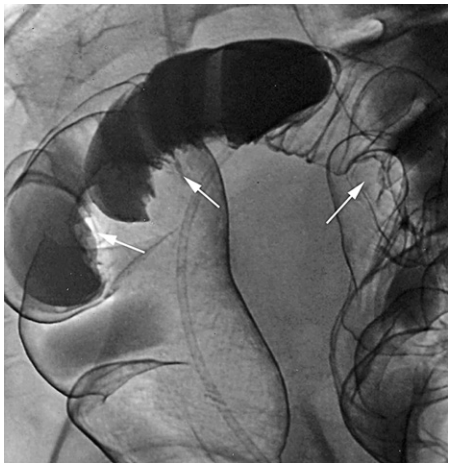




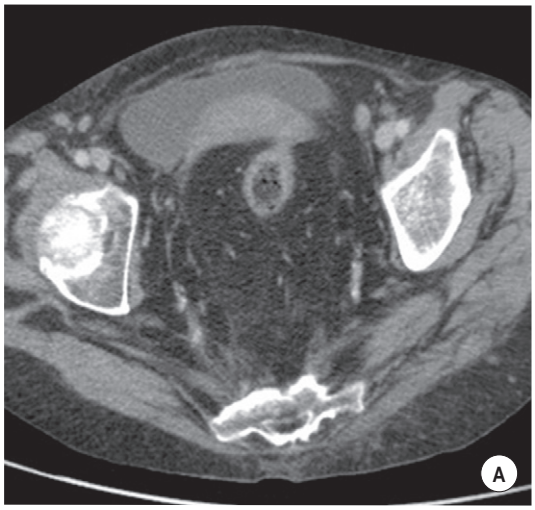
Patient with anastomotic leak following anterior resection. (A) CECT showing large pelvic collection (arrow) at the site of anastomosis. (B) Corresponding water-soluble enema showing extraluminal leak of contrast from the rectum (long arrow). There appears to be a stricture proximal to this (short arrows).

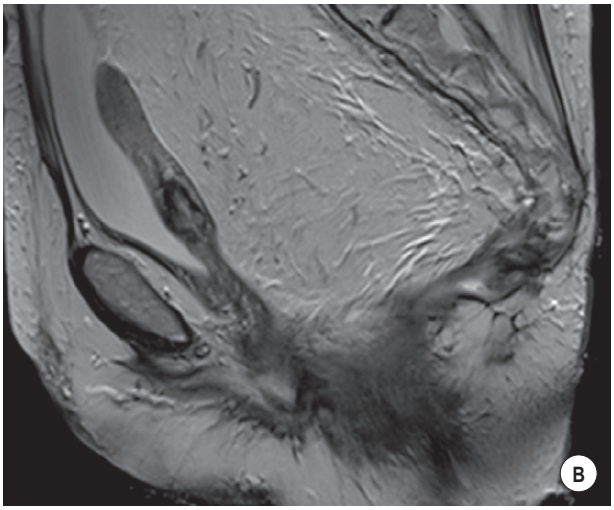


Sagittal MRI of a large tailgut cyst (arrows) with cystic and solid (C) components, the latter due to the development of a carcinoid tumour within the cyst.*



Severe pelvic endometriosis causing spiculation of the mucosa with mass defects (arrows) on the anterior wall of the rectosigmoid. The stent was required for ureteric involvement.*





Pelvic lipomatosis. (A) Axial CT and (B) sagittal T1WI demonstrating abundant pelvic fat.

3.6 LIVER

ANATOMY

Couinaud classification

- The liver is subdivided anatomically into 8 segments in an anticlockwise fashion:
  - The horizontal left and right portal veins separates the superior segments (II, IVa, VIII, VII) from the inferior segments (III, IVb, V, VI)
  - The three vertical hepatic vein branches further subdivide the segments:
    - *Right branch:* this separates segments VII, VI from VIII, V
    - *Middle branch:* this separates segments VIII, V from IVa, IVb
    - *Left branch:* this separates segments IVa, IVb from II, III
- *The caudate lobe (segment I):* this is autonomous, receiving vessels from both the left and right portal vein branches and the hepatic artery ► it has an independent venous drainage directly into the IVC

Vascular anatomy

- *Blood supply to liver:* ⅔ is from the portal vein ► ⅓ is from the hepatic artery
- *Venous drainage:* this is via the 3 hepatic veins into the IVC (30% of patients have accessory draining veins) – usually an accessory inferior RHV draining of segments VI or VII
- Aberrant gastric venous drainage of segments I and IV: this is correlated with focal fatty change within this segment

Riedel's lobe

- A normal variant where there is extension of the inferior tip of the right lobe to or beyond the costal margin

LIVER IMAGING TECHNIQUES

US

Normal texture Homogeneous (and slightly more reflective than the renal cortex)

Hepatic artery

- *Doppler:* a pulsatile vessel with continuous forward flow

Portal vein branches A radiating pattern from the porta hepatis (with reflective vessel walls)

- *Doppler:* monophasic flow is seen in a hepatopetal direction (cf. hepatofugal flow with cirrhosis) ► there is a mean peak velocity of 15–25cm/s with slight respiratory variation

Hepatic vein branches A radiating pattern from the IVC (with non-reflective vessel walls)

- *Doppler:* there is a triphasic flow pattern with reversal of flow during the cardiac cycle (reflecting transmitted right heart pressure changes)

Contrast-enhanced US This gives improved lesion characterization during the arterial and portal phases of enhancement (after an IV injection of a microbubble contrast agent)

CT

NECT This can detect diffuse changes (e.g. fat and iron deposition) and focal changes (e.g. calcification and haemorrhage) ► the liver usually has attenuation values of 54–60HU (8–10HU greater than the spleen)

CECT This can detect and characterize focal lesions using a combination of early and late arterial phase studies along with portal, late and delayed imaging

- The majority of pathological solid liver lesions have a predominantly arterial supply (normal liver parenchyma receives up to 80% of its blood supply from the portal vein) ► they will therefore appear low attenuation on portovenous imaging.

MRI

- There are a wide range of protocols available, including breath-hold T1WI and T2WI, in- and out-of-phase sequences (for fat detection), diffusion imaging, and T1WI + Gad
  - The biliary system can be imaged using a dedicated heavily T2W MRCP technique

Contrast agents

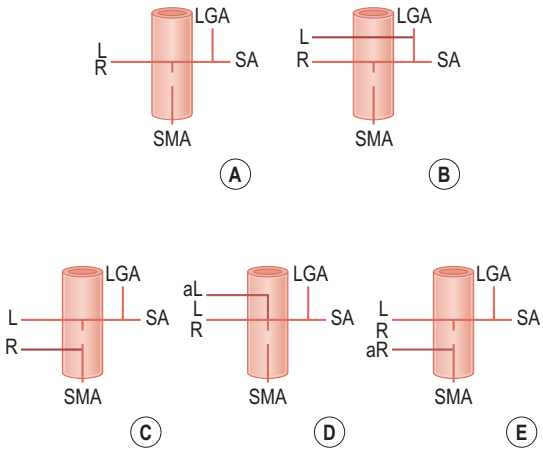
- **Gadolinium-based agents:** these will generate enhancement (T1WI)
- **Hepatobiliary specific agents**: the target includes the reticuloendothelial system or hepatocyte
  - *Iron oxide particles:* these are superparamagnetic causing susceptibility induced proton dephasing (with a reduced SI) within normal tissues on T1WI and particularly T2WI ► larger particles (50–100nm) are taken up by the Kupffer and endothelial cells and are rapidly cleared from the circulation ► smaller particles are retained within the circulation for a longer period providing a prolonged 'intravascular' phase of enhancement (and therefore providing an angiographic effect as a blood pool agent)
  - *Hepatocyte-specific paramagnetic agents* (e.g. Mn-DPDP [mangafodipir trisodium]) ► these accumulate within hepatocytes, and then undergo biliary excretion ► they will cause enhancement of normal liver parenchyma and the biliary tree (T1WI) – a low SI indicates an abnormal area

Normal imaging appearances The liver demonstrates the same (or slightly higher) SI than adjacent muscle (for all sequences except for inversion recovery techniques which are designed to null the liver signal)

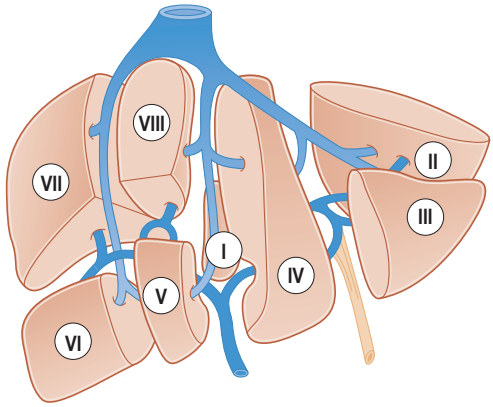
- T1WI: spleen < liver
- T2WI: spleen > liver

Liver scintigraphy

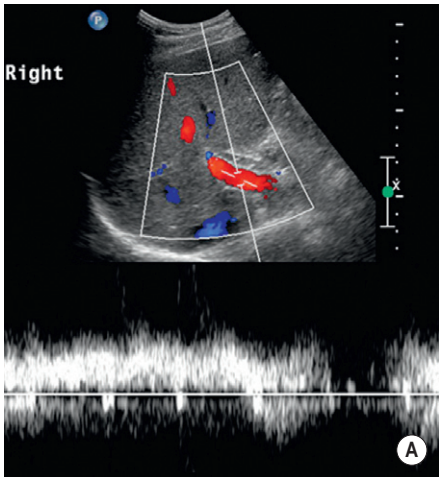
- This provides a global view of the liver and helps characterize a lesion if CT or MRI is not available
- ^{99m}Tc-sulphur colloid or albumin colloid is usually used – 90% is taken up by the Kupffer cells (10% is taken up by the spleen) ► ^{99m}Tc-labelled red blood cells can be used if a haemangioma is suspected
- FDG-PET has a relatively limited role (as normal liver takes up FDG)

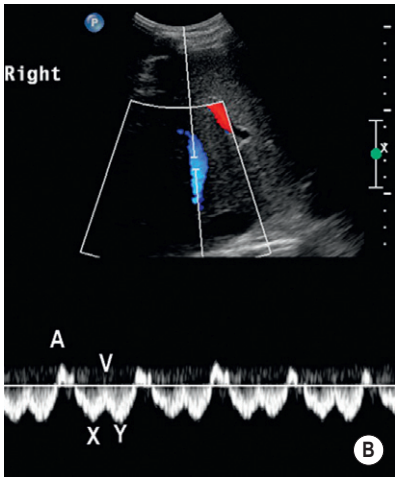


Hepatic artery normal variants. The normal arrangement is shown in (A). The commonest four variations are: replaced left hepatic artery (B), replaced right hepatic artery (C), accessory left hepatic artery (D), accessory right hepatic artery (E). R = right hepatic artery, L= left hepatic artery, LGA = left gastric artery, SMA = superior mesenteric artery, SA = splenic artery, a = accessory.

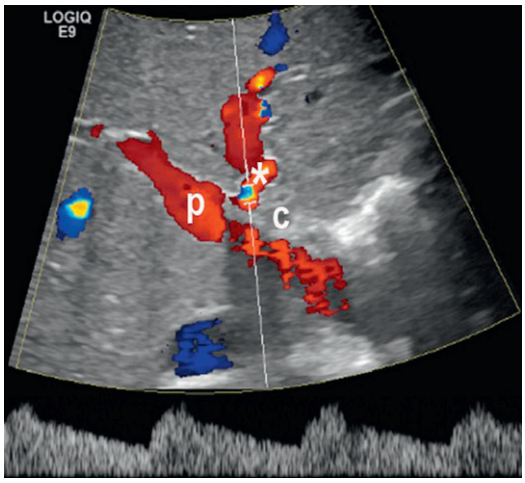


Surgical segments of the liver.

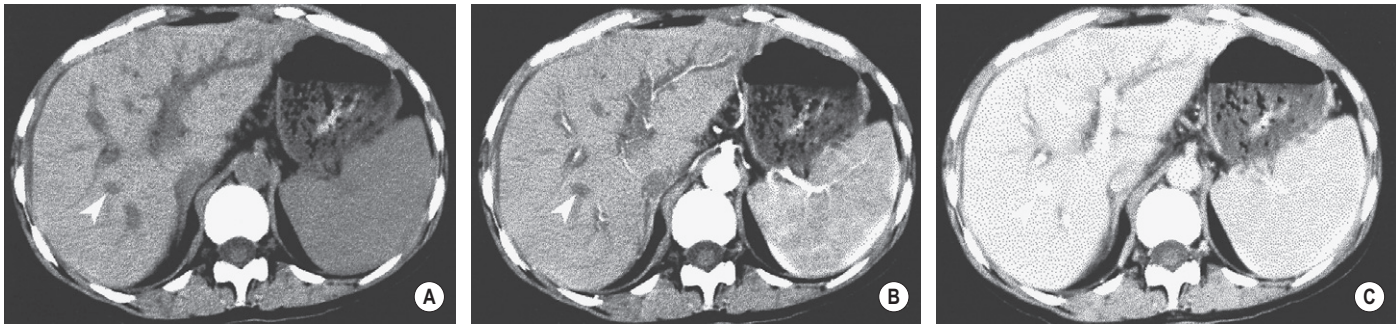




(A) Normal portal vein on duplex US. Flow is normally continuous towards the liver (hepatopetal) with slight undulation related to the cardiac cycle and respiration. (B) Normal hepatic vein on duplex US. The spectral tracing reflects the normal right heart pressure changes leading to flow reversal occurring normally during the 'A' wave (right atrial contraction) and occasionally during the 'V' wave. The 'X' and 'Y' descents are also normally demonstrated.



Normal hepatic artery (*) demonstrating pulsatile flow. p = portal vein, c = common bile duct.



Biphasic CT examination. Axial sections at the same location following a bolus of IV contrast medium demonstrating clearly the hepatic vessels and phases of enhancement, (A) NECT, (B) early arterial phase and (C) portal phase. The patient has mild intra-hepatic bile duct dilatation emphasizing the anatomical relationships. Note the hepatic vein appears as a focal lesion on the arterial phase but normally fills in during the portal phase (arrowheads).*

FAT INFILTRATION/STEATOSIS

Definition Diffuse hepatocyte triglyceride loading

- *Causes:* acute and chronic alcohol abuse ► obesity ► diabetes mellitus ► cystic fibrosis ► malnourishment ► total parenteral nutrition ► tetracyclines ► steroids ► ileal bypass surgery

US An increased echoreflectivity, which obscures the portal vein margins

CT The attenuation decreases by approximately 1.6HU per mg of triglyceride increase per gram of liver substance ► the liver architecture is preserved ► there will be uniform enhancement post IV contrast medium administration

- *Moderate fat infiltration:* liver attenuation < spleen attenuation
- *Severe fat infiltration:* liver attenuation < blood attenuation (the hepatic vasculature appears 'enhanced')

MRI Chemical shift or 'in- and out-of-phase' imaging allows for diagnosis and quantification

HEPATITIS

Definition Acute or chronic liver inflammation

- **Infection:** this is usually due to hepatitis
  - *Hepatitis A:* this is usually benign and self-limiting
  - *Hepatitis B:* this can present as an asymptomatic carrier state, or with acute or chronic hepatitis, fulminant hepatic failure, and hepatocellular carcinoma
  - *Hepatitis C:* an acute or chronic hepatitis with possible subsequent cirrhosis
- **Other causes:** alcohol ► drugs (e.g. methotrexate)

US

- *Acute hepatitis:* non-specific reduced reflectivity with echogenic portal vein walls ► gallbladder wall thickening
- *Chronic hepatitis:* an increased echogenicity with loss of the portal vein wall echogenicity

Colloid scintigraphy There are similar appearances to early cirrhosis but with uneven and reduced uptake

CT/MRI/angiography This is of limited value until cirrhosis develops

HAEMOCHROMATOSIS AND HAEMOSIDEROSIS

Haemochromatosis

- An autosomal recessive condition causing iron deposition within the hepatocytes (leading to subsequent cirrhosis) and other organ tissues (including the myocardium, skin and pancreas)
  - *There is an increased risk both of developing a malignancy in general (and HCC in particular)*

Haemosiderosis

- This is due to hepatic iron overload resulting from multiple transfusions ► uptake occurs via the reticuloendothelial system (e.g. the Kupffer cells within the liver, bone marrow and spleen)
  - *There is less risk of liver damage*

US This may demonstrate increased parenchymal echogenicity

NECT Increased liver attenuation values (HU>75) ► previous amiodarone treatment and Thorotrast exposure may give similar appearances

MRI This is the most specific imaging technique ► intracellular iron deposition exerts a local susceptibility effect leading to an abnormal reduction in liver SI ► NB: normal hepatic parenchyma is brighter than adjacent skeletal muscle on T1W1 + T2W1

- It is best detected with T2* gradient-echo sequences (moderate accumulation will cause changes on T2WI – severe accumulation will cause changes on T1WI)
- *Haemochromatosis:* reduced SI in the liver, pancreas and heart
- *Haemosiderosis:* reduced SI in the liver and spleen

NEONATAL HEPATITIS

Causes *Idiopathic* (the majority) ► *antenatal infections* (e.g. CMV, rubella, enterovirus, toxoplasmosis, herpes simplex, and spirochaete) ► *metabolic disorders* (e.g. cystic fibrosis, α_1-antitrypsin deficiency, tyrosinaemia and galactosaemia)

US There are non-specific features, but may include: hepatomegaly ► a heterogeneous coarse liver parenchyma ► a visible gallbladder (>1.5cm) without the triangular cord sign (cf. biliary atresia)

^{99m}Tc-DISIDA Extraction into the liver is often reduced and excretion into the bowel may be reduced proportional to the degree of cholestasis and hepatocellular dysfunction ► if cholestasis is severe, reduced extraction and excretion may make it difficult to distinguish this from biliary atresia

Pearls

- 5–10% will develop persistent fibrosis
- Diagnosis: percutaneous liver biopsy

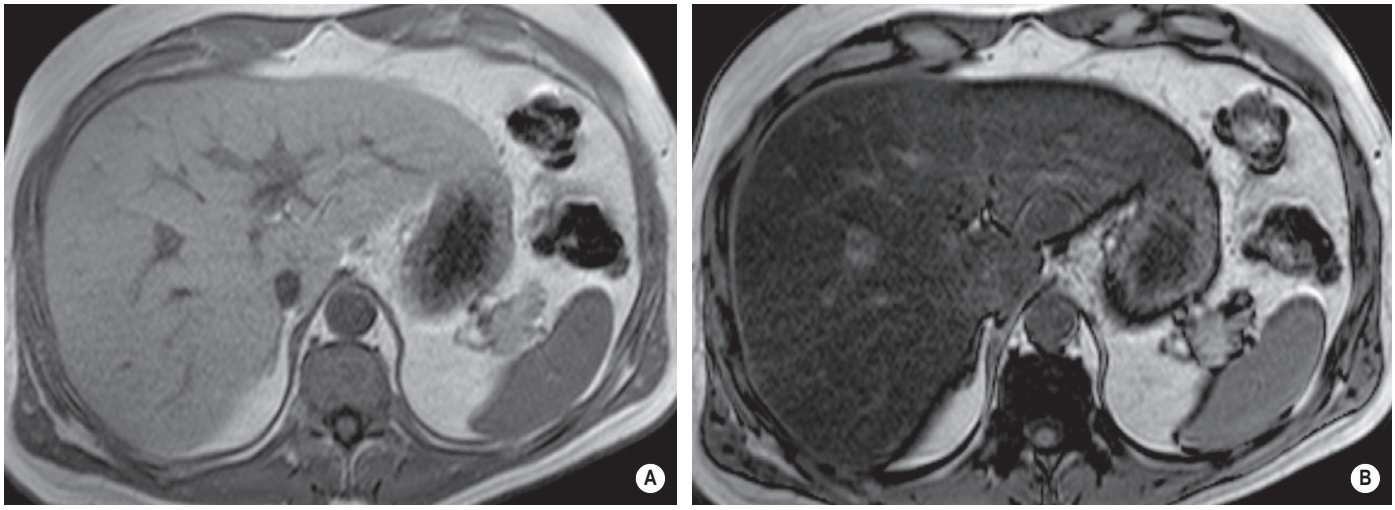
WILSON'S DISEASE

Definition An autosomal recessive condition causing copper deposition within the liver, cornea and lenticular nucleus of brain ► this is hepatotoxic and triggers an inflammatory response that progresses to cirrhosis

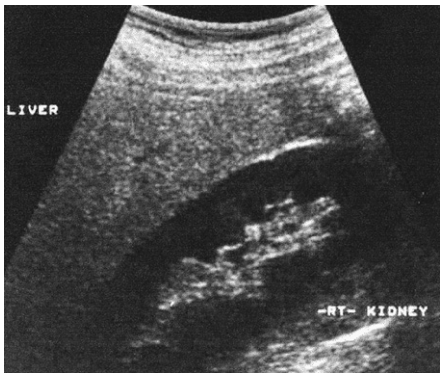
US Non-specific features ► generalized cirrhotic changes

CT There is rarely increased hepatic attenuation (more often there is reduced attenuation secondary to fatty infiltration)

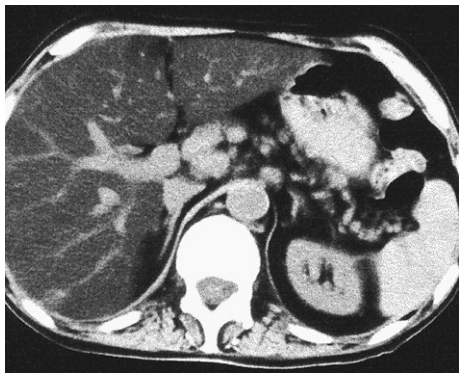
MRI T1WI: possibly high SI ► T2WI: low SI



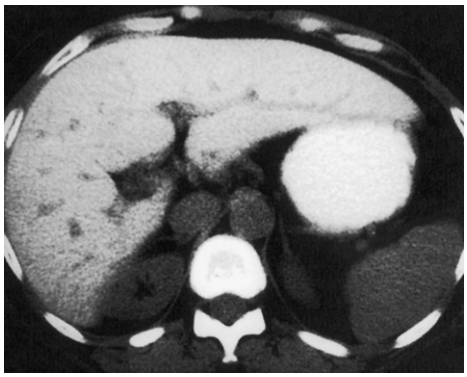
Liver steatosis. (A) In-phase imaging. (B) Out-of-phase imaging. The signal loss demonstrated by the hepatic parenchyma on out-of-phase imaging indicates significant liver steatosis.



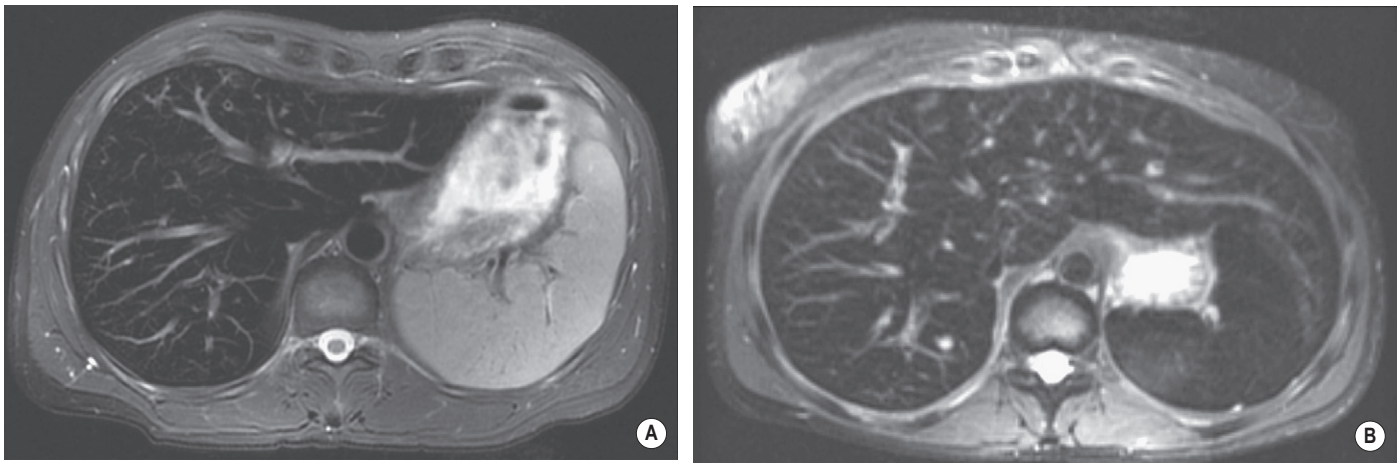
Diffuse fat infiltration. The liver is abnormally echoreflective when compared with the cortex of the adjacent right kidney.*



Diffuse fat infiltration. NECT in which the liver parenchyma is markedly reduced in attenuation, reversing the normal relationship with the spleen and blood vessels. The shape and vascular architecture of the liver are normal.*



Haemochromatosis. NECT section through the liver of a patient with haemochromatosis showing diffuse increased attenuation of the liver compared with the spleen.*



T2WI images demonstrating abnormally low liver signal in (A) haemochromatosis and (B) haemosiderosis. Note the spleen is abnormally low signal in haemosiderosis as the reticuloendothelial system becomes iron loaded compared with predominantly hepatocyte accumulation in haemochromatosis.*

CIRRHOSIS

DEFINITION

- This represents the endpoint of a wide variety of chronic disease processes that cause hepatocellular necrosis and ultimately lead to hepatic fibrosis and nodular regeneration:
  - Alcohol abuse ► hepatitis B ► storage disorders (e.g. haemochromatosis and Wilson's disease) ► biliary cirrhosis ► certain drugs

RADIOLOGICAL FEATURES

US

- *Early cirrhosis:* there is increased reflectivity (due to fat infiltration and fibrosis)
- *Advanced cirrhosis:* a nodular liver margin (this is seen especially with a high-frequency transducer and if ascites is present) ► a coarse heterogeneous echotexture ► hypoechoic regenerating nodules
  - Attenuated hepatic veins can be seen with end-stage disease (due to the liver atrophy)
  - Pure hepatic fibrosis increases liver reflectivity (resulting in loss of the portal vein branch margins) but will not significantly alter its attenuation – this can be used to discriminate fibrosis from fatty infiltration

Doppler US Damping of the normal right-heart waveforms within the hepatic veins ► reduced main portal venous blood flow (<10cms^{-1} mean peak) or hepatofugal portal venous flow ► collateral vessel development (e.g. left gastric, splenorenal, paraoesophageal, or retroperitoneal collaterals) including a recanalized para-umbilical vein

- Increased hepatic arterial flow can be seen with advanced cirrhosis (due to a reduced portal venous contribution to the hepatic blood supply)
- Increased flow in a large recanalized para-umbilical vein may 'steal' blood from the right portal vein branch – this can lead to reversed flow within the right portal vein but normal hepatopetal flow within the main and left portal veins

CT This is relatively insensitive for early cirrhosis

- *Advanced cirrhosis:* a nodular liver margin ► lobar atrophy or hypertrophy ► ascites ► portal vein thrombosis
  - Heterogeneous attenuation: this is often due to coexisting fibrosis (with reduced attenuation) and hepatocyte iron deposition (with increased attenuation)

MRI This is relatively insensitive for early cirrhosis

- *Early cirrhosis:* T2WI and delayed T1WI + Gad: subtle parenchymal heterogeneity
- *Advanced cirrhosis:* morphological changes as seen with CT ► it can assess portal vein patency, flow direction and bulk flow volume

Colloid scintigraphy

- *Early cirrhosis:* there is uneven radionuclide uptake (and lobar morphological changes with progression)
  - *'Colloid shift':* with the development of portal hypertension there is splenomegaly and a reduced activity halo around the liver
- *Advanced cirrhosis:* less sulphur colloid is taken up by the liver and increased extrahepatic activity is seen within the heart, and the reticuloendothelial cells of the bones and lungs

Angiography

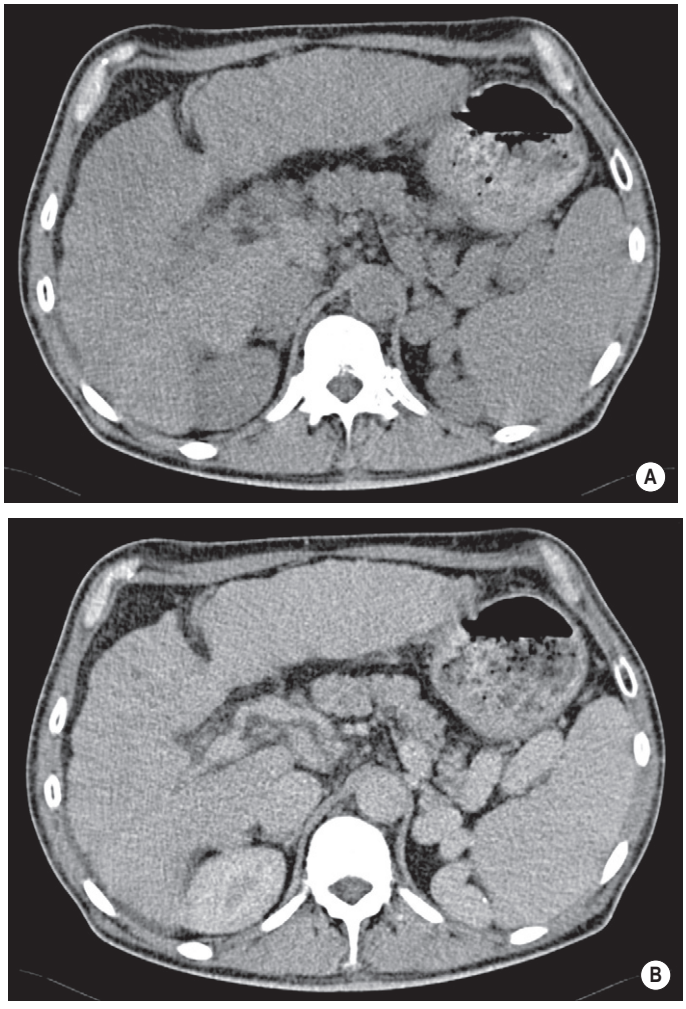
- This can assess any vascular complications and portal hypertensive changes
- *Hepatic arteriography:* there is increased tortuosity of the intrahepatic branches ('corkscrew vessels'), reflecting hepatic lobar shrinkage

PEARLS

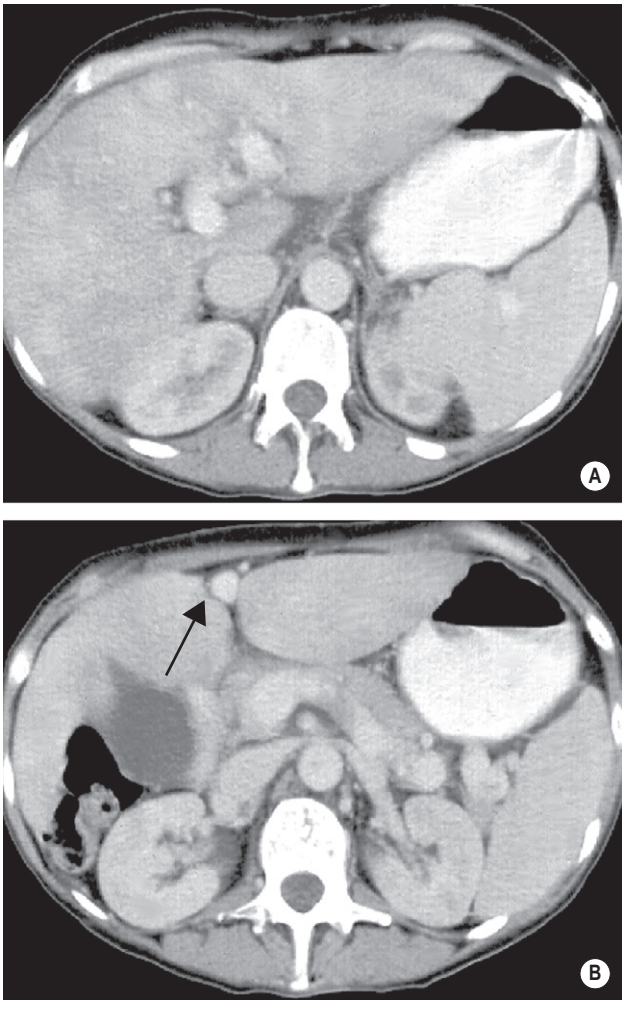
Advanced cirrhosis There is atrophy of the posterior segments of the right lobe (VI and VII), hypertrophy of the caudate lobe (I) and hypertrophy of the lateral segments of the left lobe (II and III)

- This is thought to be caused by altered hepatic blood flow dynamics, including an increased overall hepatic blood flow (due to intrahepatic arteriovenous shunts) and areas of reduced hepatic blood flow (due to increased intrahepatic vascular resistance)

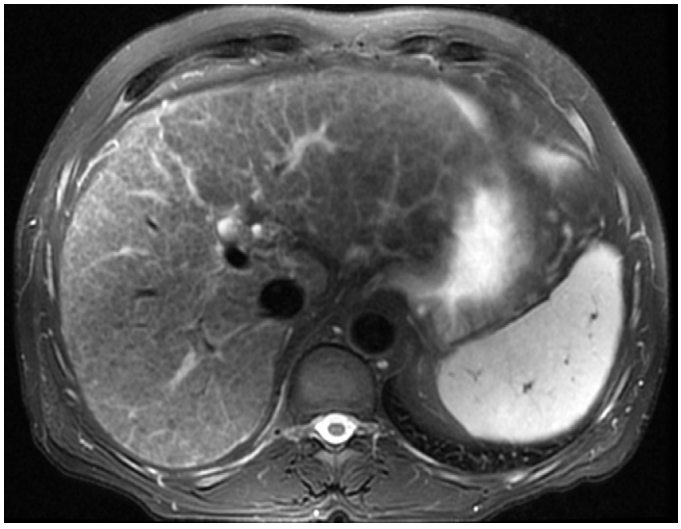
Complications Hepatocellular carcinoma (10%) ► portal hypertension (± variceal bleeding)



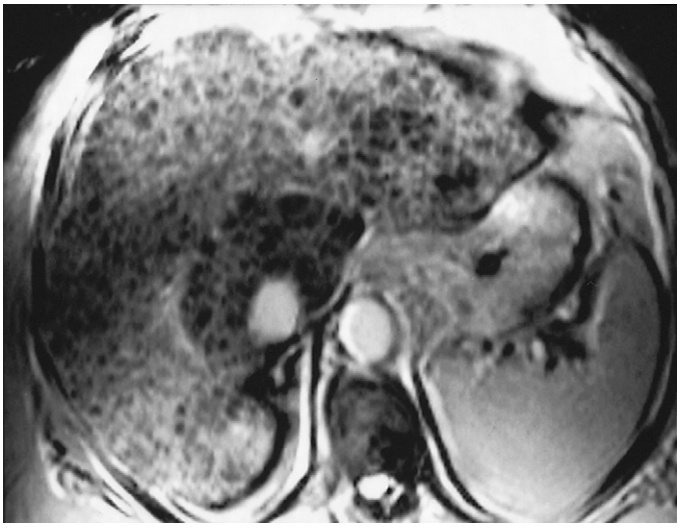
Cirrhosis. On (A) NECT (B) portal phase CECT the nodular margin, atrophy of the hepatic right lobe and large splenorenal varices are all indicators of cirrhosis.*



CT appearances of cirrhosis (A, B) Patchy irregular enhancement of the hepatic parenchyma due to altered portal blood flow in cirrhosis with portal hypertension. Recanalization of the umbilical vein in the falciform ligament is arrowed.†



Cirrhosis. On T2WI imaging the marked heterogeneity may be apparent in a cirrhotic liver, due to the combination of increased signal from fibrosis and reduced signal from iron accumulation within nodules.*



Cirrhosis. SPIO-enhanced T2 image illustrates the nodular architecture of the cirrhotic liver. Nodules of regenerating liver tissue show SPIO uptake giving low signal, while interstitial bands of fibrosis show relatively high signal.†

BENIGN SOLID LIVER LESIONS

HAEMANGIOMA

Definition

- These are composed of vascular channels of varying size (cavernous to capillary) which are endothelium lined ► there is often intervening fibrous tissue of varying amounts
  - *Capillary haemangioma:* the usual form
  - *Cavernous haemangioma:* this accounts for most neonatal and infantile haemangiomas (and some adult lesions)
- It is the commonest benign hepatic tumour (it can be multiple in <10% of cases)

Clinical presentation

- They are usually asymptomatic ► larger lesions may rarely cause discomfort or undergo spontaneous rupture (F>M)
- They may enlarge during pregnancy

Radiological features

US

- *Capillary haemangioma:* a well-defined lobular homogeneous hyperechoic lesion (large lesions can be heterogeneous) ► there is no Doppler signal (due to the very slow vascular flow through the dilated channels) ► it can appear very similar to some metastases (e.g. from a GI primary)
- *Cavernous haemangioma:* a hypoechoic lesion (due to the larger vascular channels) ► a Doppler signal is usually detectable (there are more rapid flow rates)

CT A well-defined lobulated lesion ► thrombi, calcification, fibrosis and scarring are variably present

- *NECT:* there is a similar attenuation to blood
- *CECT:* there is centripetal enhancement – the lesion eventually merges with the background parenchyma

MRI A well-defined lobulated lesion ► characteristic imaging features are demonstrated if a lesion is between 2 and 4cm in size.

- *T2WI:* there is increasingly high SI with extended echo times (malignant lesions are typically less prominent with later echo times)
  - *'Lightbulb' sign:* homogeneously high SI (greater than that of the spleen and approaching that of cyst fluid)
- *T1WI + Gad:* centripetal enhancement from the periphery to the centre over a period of minutes ► there are three distinct enhancement patterns:
  1. A well-circumscribed hepatic mass with peripheral, nodular and interrupted enhancement progressing centripetally to uniform enhancement (most common)
  2. Immediate uniform enhancement (small capillary haemangiomas <1.5cm).
     - This will also show persistent delayed enhancement
  3. Peripheral nodular enhancement with centripetal progression but persistent central hypointensity (giant haemangiomas >5cm).
  - *Small (<1.5cm) lesions:* these may fail to demonstrate characteristic T2WI signal changes (due to partial volume effects) or the typical enhancement pattern
  - *Larger (>4cm) lesions:* these often have atypical internal features such as an area of central fibrosis that can prevent complete infilling during contrast enhancement
- *DWI:* hyperintense (T2 shine through)

Sulphur colloid studies Lesions appear as photopenic areas

Blood pool studies (e.g. ^{99m}Tc-labelled red cells) Lesions demonstrate increased uptake

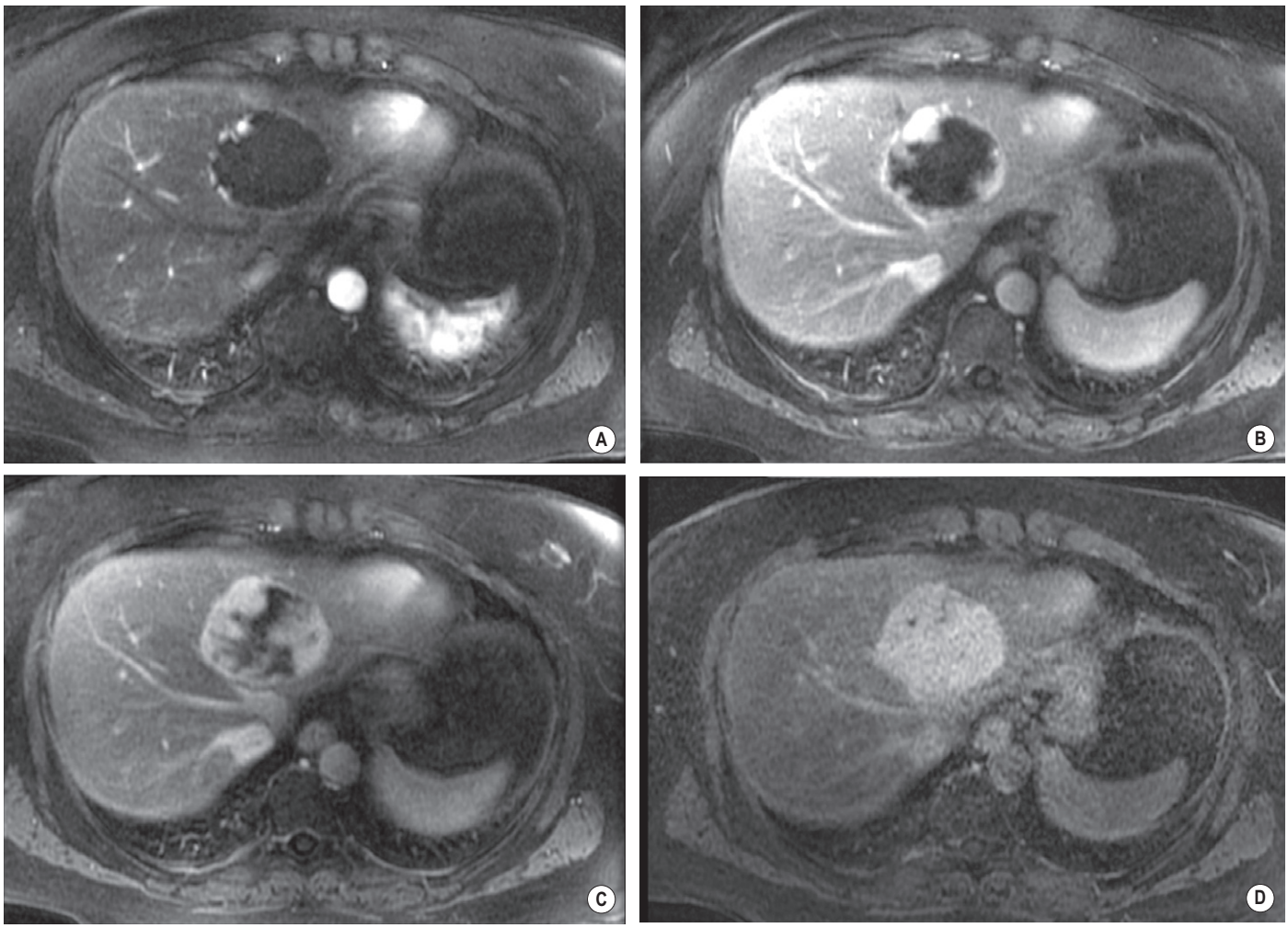
DSA A characteristic 'cotton wool' appearance: normal-sized arteries supply groups of peripherally arranged vascular spaces that opacify gradually and retain contrast for 20 s or longer

Pearls

- Haemangiomas are more T2 hyperintense than are most metastases, although hypervascular metastases can mimic haemangiomas because of their marked T2 hyperintensity
  - Delayed (>5 min) contrast-enhanced images are helpful in these cases because small, uniformly enhancing haemangiomas retain contrast material and remain hyperintense, whereas hypervascular metastases will show 'washout' of contrast material

Diagnosis This may require a core needle biopsy in adults if the diagnosis is in doubt

Kasabach–Merritt syndrome A large haemangioma may sequester thrombocytes, leading to a thrombocytopenia



Haemangioma. Typical appearance of enhancement following IV gadolinium with initial peripheral nodular high signal vessel signal followed by progressive infilling of the lesion. Images obtained at 40 s (A), 120 s (B), 5 min (C) and 15 min (D) following injection.*

Typical imaging features of common liver lesions

	Adenoma	FNH	Haemangioma	HCC*	FLC**
Sex	F > M	F > M	F > M	M > F	M = F
Capsule	Yes	No	No	Thin	Thin
Central scar	Uncommon	Yes High SI (T2WI)	No	Uncommon	Yes Low SI (T1WI and T2WI)
Calcification	No	No	Yes	7%	50%
Enhancement	Uniform arterial	Uniform arterial ► delayed scar enhancement	Centripetal	Arterial (may be mosaic) ► portovenous washout	Uniform arterial ► there is no scar enhancement
Colloid scintigraphy	Reduced uptake	Normal uptake	Reduced uptake	Reduced uptake in less well-differentiated tumours	Reduced uptake

FNH, focal nodular hyperplasia
*Hepatocellular carcinoma
**Fibrolamellar carcinoma

FOCAL NODULAR HYPERPLASIA (FNH)

Definition

- An underlying congenital vascular lesion composed of normal liver elements (hepatocytes, bile ducts, Kupffer cells and intervening fibrous septa) ► however, there is a lack of normal liver architecture (e.g. there are absent portal tracts)
  - It may enlarge in response to hormone stimulation (e.g. oral contraceptives)
- It is the 2nd commonest benign hepatic tumour

Clinical presentation

- It is usually asymptomatic (it may present with pain or hepatomegaly)
- It occurs most commonly in women aged 20–50 years (and is multiple in 20% of cases)

Radiological features

- A central stellate fibrovascular scar is seen in 50% of cases ► there is no true capsule ► calcification, necrosis and haemorrhage are extremely rare (even large lesions do not usually outgrow their blood supply)

US There are non-specific features with lesions demonstrating a similar reflectivity to the adjacent liver (but demonstrating mass effect) ► the central scar is rarely seen

- Doppler signals can be seen within and at the edge of the lesion

NECT A well-defined mass which often exhibits a mass effect (with vessel displacement) ► the lesion demonstrates the same attenuation as the surrounding liver ► there is a central low attenuation scar

CECT

- *Arterial phase:* uniform enhancement (except for the scar) ► there can be large peripheral feeding vessels
- *Portal phase:* the attenuation is identical to normal liver (the scar remains low attenuation)
- *Delayed imaging:* there is slow scar enhancement

MRI The same enhancement pattern is seen as for CT ► the specificity increases with iron oxide agents (which are taken up by the Kupffer cells)

- *T1WI:* intermediate or minimal low SI ► a low SI central scar
- *T2WI:* intermediate to high SI ► a high SI central scar
- *T1WI + Gad:* marked, homogeneous arterial phase enhancement that becomes isointense during the portal venous phase ► there can also be a peripheral, ring-type delayed enhancement pattern on delayed images obtained 1 h after hepatocyte selective gadolinium chelate administration
  - The central scar usually demonstrates delayed enhancement
- *DWI:* generally isointense

Sulphur colloid This is usually normal (due to Kupffer cell activity within the lesion)

DSA A vascular mass with a large tortuous central supplying artery ► radiating vessels spread out to supply the lesion

Pearls

- *Other lesions with a central scar:* hepatocellular adenoma (HCA) ► hepatocellular carcinoma ► haemangioma
- *Other lesions demonstrating Kupffer cell activity:* HCA ► a well-differentiated hepatocellular carcinoma
- MR imaging with a hepatocyte-specific contrast agent may help confirm the hepatocellular origin of the mass:
  - MultiHance (Gd-BOPTA): iso- to hyperintense on 1-h to 3-h delayed images, unlike an adenoma (which is hypointense)

FOCAL CONFLUENT FIBROSIS

Definition

- A focal manifestation composed of massive confluent fibrosis and associated atrophy of the affected liver segment
- It occurs uncommonly in established cirrhosis, typically affecting the anterior segment of the right lobe or the medial segment of the left lobe

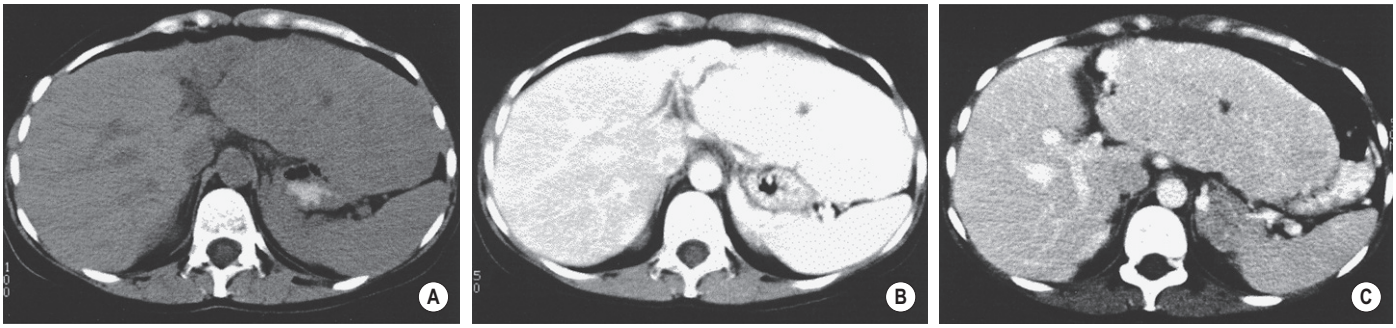
Radiological features

NECT The involved atrophic segment is of low attenuation (often with retraction of the overlying capsule)

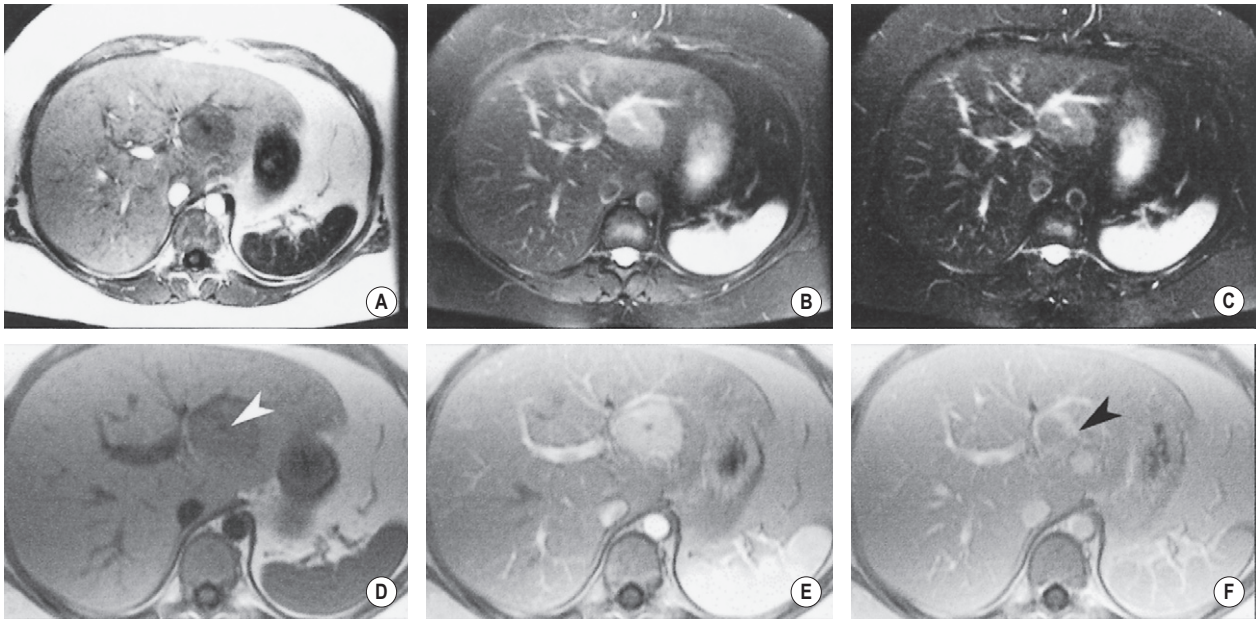
CECT The attenuation of the affected region is the same or lower than normal liver

MRI Similar morphological changes are evident

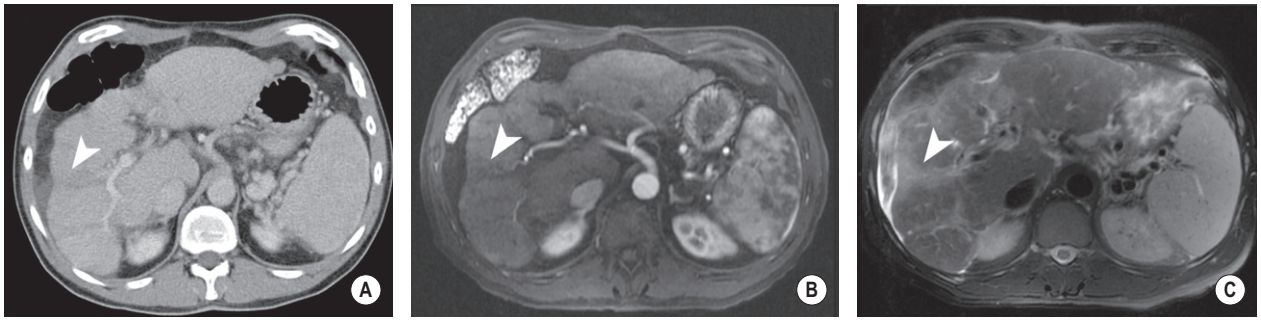
- *T1WI:* low SI
- *T2WI:* high SI
- *T1WI + Gad:* delayed enhancement



FNH. A large lesion (homogeneous except for a low attenuation central scar) expands and replaces much of the left lobe. The attenuation is almost identical to normal liver on the unenhanced section (A) but the lesion demonstrates transient avid and even enhancement during the arterial phase (B), becoming indistinguishable on the portal phase section (C).*



FNH. A well-defined lesion in the left lobe that is homogeneous except for a central scar, low signal on T1WI (A, D) and increased signal on T2WI (TE 60 ms (B), TE 120 ms (C)). Following IV gadolinium the whole lesion enhances transiently in the arterial phase (E) with the exception of the scar, which enhances on delayed sections (F) (arrowhead).*



Focal confluent fibrosis. (A) Portal phase CECT. The right lobe has contracted and confluent bands of fibrosis are present (arrowheads). These appear of low attenuation and may mimic malignant lesions when focal. (B) On T1WI + Gad imaging there is no enhancement. (C) On T2WI the fibrosis is usually of increased signal and can mimic HCC.

HEPATIC ADENOMA

Definition

- A rare benign hepatic tumour arising spontaneously or in association with glycogen storage disease type 1 (when it is often multiple)
- It is a vascular lesion composed primarily of hepatocytes which have a tendency to accumulate fat and glycogen ► there are no portal tracts or bile ducts present ► although Kupffer cells are absent, Kupffer cell activity can be seen in up to 20%
- It may demonstrate a fibrous pseudocapsule and a central scar (like FNH) ► there is a tendency to outgrow its blood supply, resulting in haemorrhage, thrombosis and necrosis
  - *Aetiological factors:* oral contraceptives ► androgenic steroids

Clinical presentation

- It is usually asymptomatic ► if rupture occurs there can be pain or life-threatening haemorrhage

Radiological features

US An isoechoic or hyperechoic (if there is a significant fat component) mass lesion

CT Uncomplicated lesions are usually homogeneous with a well-defined margin ► it will be of similar attenuation to normal liver (or lower attenuation if there is a substantial fat component) ► intraparenchymal haemorrhage will appear as high attenuation thrombus on unenhanced images (which may extend through the capsule and into the peritoneum)

- *Arterial phase:* there is marked uniform enhancement
- *Portal phase:* a lesion merges with the surrounding liver
  - Necrosis: minimally enhancing low attenuation regions

MRI An uncomplicated adenoma has similar appearances to a region of FNH

- T1WI: a well-defined isointense or slightly high SI lesion ► there can be hyperintense foci secondary to haemorrhage or intracellular fat
- T2WI: variable signal intensity but are often mildly hyperintense relative to the liver ► haemorrhage or necrosis: this leads to a heterogeneous appearance
- T1WI + Gad: heterogeneous or uniform marked enhancement (arterial phase), equilibrating with the liver parenchyma (portal phase) ► they are typically not as vascular as FNH
  - They can demonstrate delayed contrast material washout with or without a delayed-enhancing pseudocapsule
  - They are hypointense on 1-h to 3-h delayed images obtained with Gd-BOPTA
  - Although venous washout is generally worrisome for malignancy, an adenoma may demonstrate venous washout (usually worrying for malignancy) – it is the only 'benign' hypervascular mass that may do so (cf. delayed isoenhancement with FNH)
- DWI: variable signal intensity depending on the presence of blood or necrosis

Sulphur colloid Reduced activity

HIDA There is uptake within a lesion but no excretion (due to the absent bile ducts)

DSA The lesion is usually hypervascular

Pearls

- As there is a 1% risk of malignant change, resection is usually preferred to conservative management
- **Liver adenomatosis:** multiple, progressive, symptomatic adenomas (these are not steroid dependent) ► there is a risk of liver dysfunction, haemorrhage and HCC formation

ATYPICAL REGENERATIVE NODULES

Definition

- These occur with chronic cirrhosis ► some regenerative nodules may appear more prominent than others, causing diagnostic confusion with a HCC
- Form part of the spectrum of HCC development (regenerative nodule → dysplastic nodule → HCC)

Radiological features

US A well-defined, homogeneous lesion of reduced reflectivity

T1WI Iso- to hyperintense (high SI due to fat and glycogen accumulation)

T2WI Iso- to hypointense ► low SI if there is accumulation of iron ('siderotic nodules')

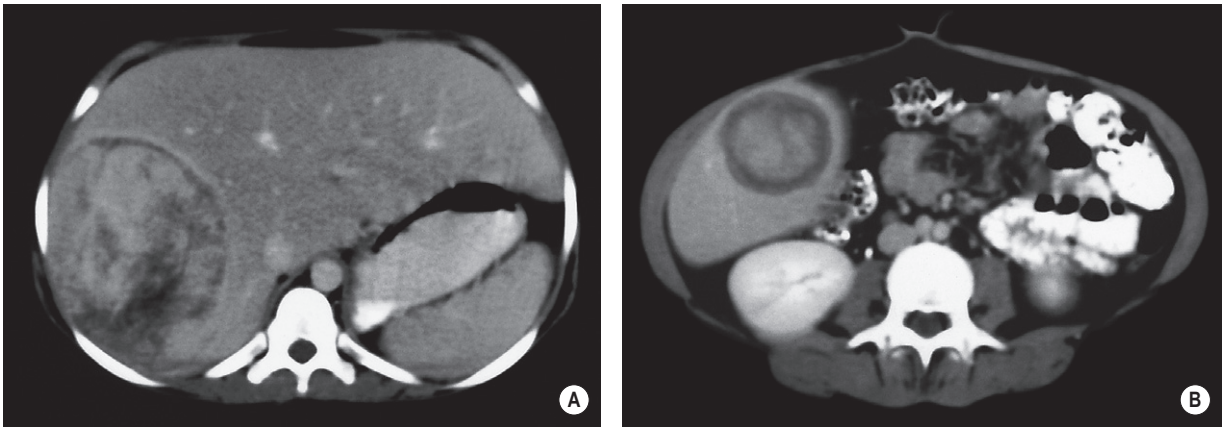
- A malignant focus within a nodule appears as a region of high SI

T1WI + Gad Multiple small, similar-sized enhancing nodules are demonstrated during the hepatic arterial phase ► these nodules then fade to isointensity (differentiating from HCC)

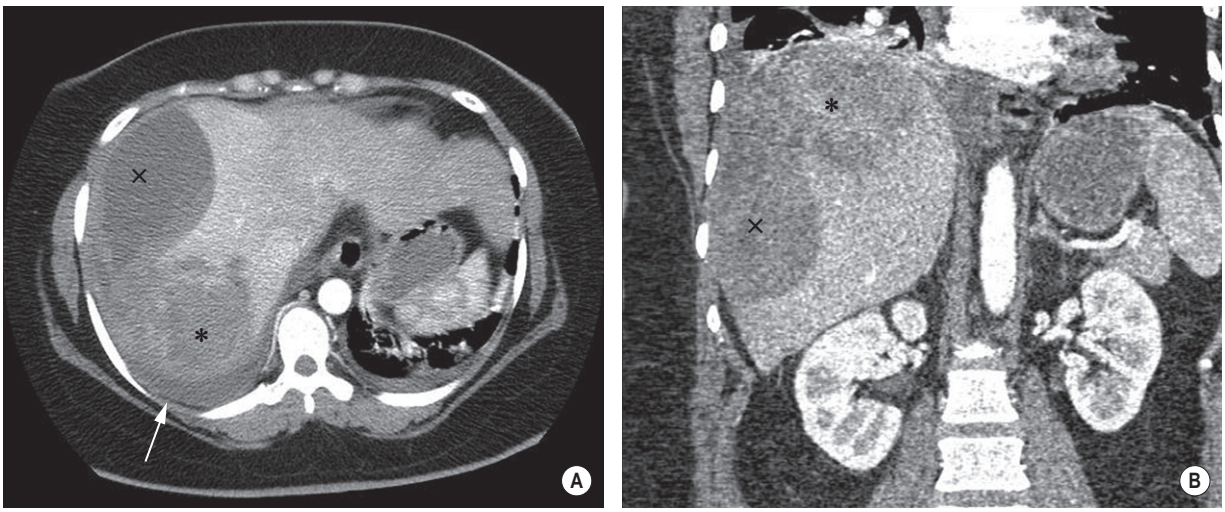
DWI Generally isointense

Pearl

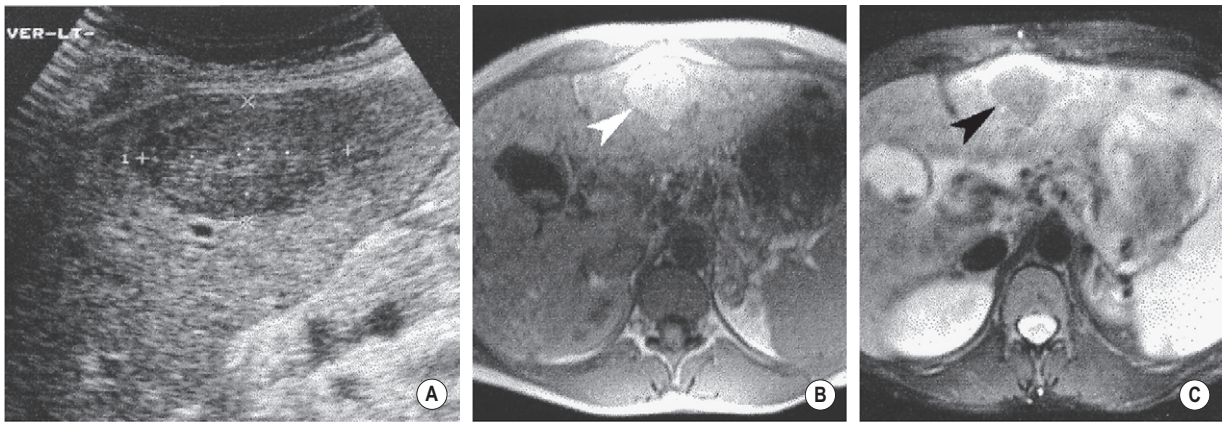
- Increasing SI on T2WI or increasing washout on T1WI + Gad is worrying for developing dysplasia



Hepatic adenoma. A 16-year-old girl with glycogen storage disease type I. Axial CECT images through the liver show a large heterogeneous adenoma in the right lobe (A) and a smaller adenoma in the lower aspect of the right lobe (B).*



(A) CECT demonstrating a large heterogeneous adenoma (asterisk) involving the right liver lobe with associated perihepatic bleeding (arrow) as well as a subcapsular haematoma (cross). (B) A sagittal reconstruction in a different patient shows a large adenoma (asterisk) also with subcapsular bleeding (cross).



Atypical regenerative nodule. Regeneration in cirrhosis often results in heterogeneity of the parenchyma. Occasionally nodules may become large, 'atypical' or 'dominant' as in this patient. US (A) demonstrates a reduced echoreflectivity lesion in the left lobe, initially interpreted as a probable tumour. However, on MRI, the relatively homogeneous lesion (arrowheads) is of increased signal on T1WI (B) and decreased signal on T2WI (C) (arrowhead).

FOCAL FAT

DEFINITION

- Focal fat variation within the liver parenchyma is due to alterations in the underlying blood supply and venous drainage ► it can cause diagnostic confusion with a tumour
  - *Common sites:* either side of the falciform ligament ► the cranial aspect of the gallbladder fossa ► the posterior aspect of segment IV

RADIOLOGICAL FEATURES

MRI – 'chemical shift' or 'in- and out-of-phase' imaging This detects the presence of fat and water within the same image voxel ► fat and water protons have different resonant frequencies – over time these will alternatively be in and out of phase with each other ► imaging at specific predetermined times will give either in- or out-of-phase images (they are out of phase 2.2 ms after an excitation pulse and in phase 4.4 ms after excitation)

- Water and fat signal intensities will combine on the in-phase imaging, but cancel out on the out-of-phase imaging ► as both image sets use a different TE, one needs to compare any signal change with a non-fat-containing organ (e.g. spleen) or correct for T2 signal changes using T2 mapping
- Lesions containing significant amounts of fat will lose SI on the out-of-phase images (relative to the in-phase images)
- *Out-of-phase images:* these can be identified as the intra-abdominal viscera are outlined by an 'inky black' line ► this occurs because at the organ–intra-abdominal fat interface the imaged voxel contains both fat and water and will therefore lose signal intensity (voxels located internally within the organ or intra-abdominal fat will tend to contain predominantly fat or water only and therefore not lose signal intensity)

NECT/US A large area of regional fat variation has a geographic appearance with a lack of mass effect and preservation of the vascular architecture

BILIARY HAMARTOMAS

DEFINITION

- A rare benign malformation of the bile ducts ('von Meyenburg complexes')
- Typically they are small lesions (3–5mm) with a combination of solid and cystic elements ► diagnosis usually requires biopsy

RADIOLOGICAL FEATURES

US If multiple they often range from 1 to 3mm in size and are often interpreted as diffuse malignant infiltration

CT There can be cystic or solid components ► they will enhance (but remain low attenuation on unenhanced and portal phases)

MRI T1WI: low SI ► T2WI: there is a characteristic appearance of multiple high SI lesions

PEARL

- They may be indistinguishable from small metastases on US and CT and are often the cause of 'indeterminate' or 'too small to characterize' lesions

MESENCHYMAL HAMARTOMA

DEFINITION

- This is a lesion containing a mixture of bile ducts and mesenchyme
- Although rare, it is the 2nd commonest benign liver tumour or developmental lesion occurring in children

CLINICAL PRESENTATION

- It is usually seen at <2 years of age (with a peak incidence at 15–22 months)

RADIOLOGICAL FEATURES

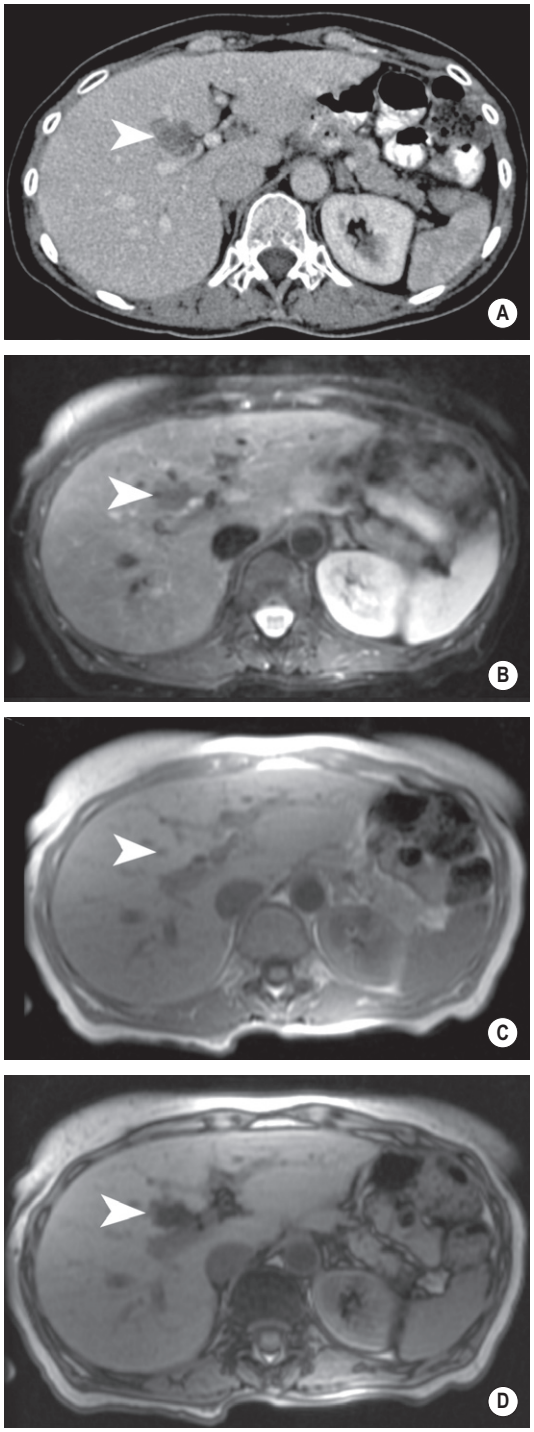
- It is usually a large lesion (5–30cm) ► it can appear as a mixed solid or cystic mass (appearing more solid when small) ► it can be multiseptated with a cystic and gelatinous composition

CT A low attenuation lesion ► there is variable septation with fluid loculation

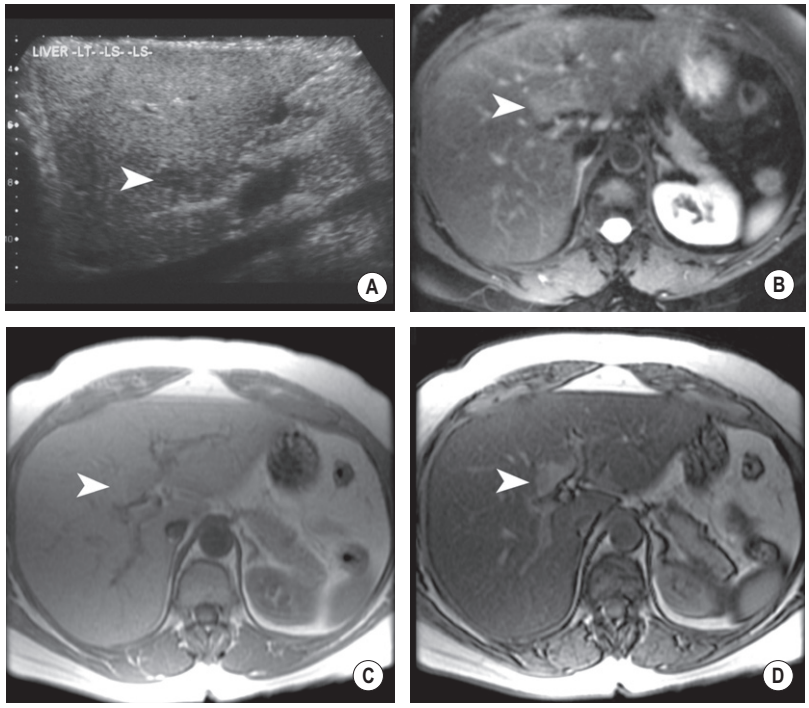
- Although the tumour is hypovascular, atrioventricular shunting can occur through the enlarged irregular tortuous feeding vessels

MRI Multiseptated fluid areas ► T1WI: low SI ► T2WI: high SI

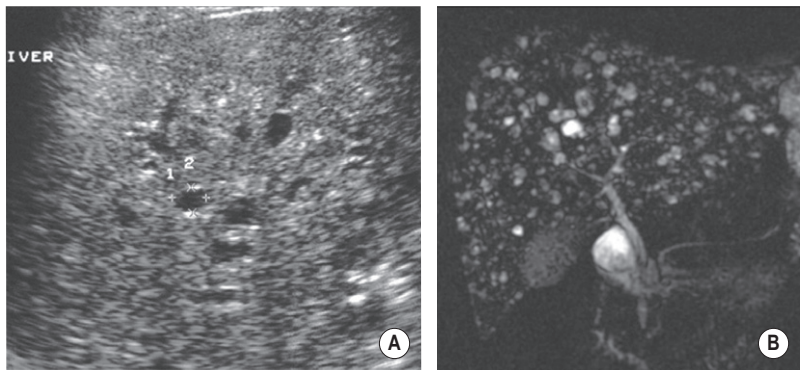
- A lesion can displace major vessels



Focal fat infiltration. On portal phase CT (A) focal fat infiltration (arrowheads) in the posterior aspect of the left lobe medial segment (Couinaud IV) has a similar appearance to a metastasis. On T2WI FS (B), the low signal makes metastasis unlikely but does not characterize the lesion. However the demonstration of signal loss (relative to the spleen) on in- (C) and out- (D) -of-phase gradient-echo imaging is diagnostic.*



Focal fat sparing. On US (A) an area of focal fat sparing (arrowheads) in the posterior aspect of the left lobe medial segment (Couinaud IV) in an otherwise fatty liver has a similar appearance to a metastasis. On T2WI FS (B) this will appear of increased signal, suggesting a malignant lesion but the use of in- (C) and out- (D) -of-phase gradient-echo imaging indicates that the 'lesion' is in fact normal liver surrounded by fatty liver that has reduced in signal on the out-of-phase image (D).*



Multiple biliary hamartomas. On US (A) only the larger cystic lesions are seen clearly and the background texture of the liver is heterogeneous and often misinterpreted as malignant infiltration. With MRCP type T2WI (B) imaging the extent and number of the multiple cystic lesions is more obvious. The solid components may be indistinguishable from metastases.*

HEPATOCELLULAR CARCINOMA (HCC) / HEPATOMA

DEFINITION

- The commonest primary malignant neoplasm of the liver, typically occurring within an abnormal (e.g. cirrhotic) liver
  - *Risk factors:* direct carcinogens (e.g. aflatoxin) ► chronic hepatitis B and C ► cirrhosis (particularly postnecrotic cirrhosis and haemochromatosis)
  - *Types:* solitary ► multifocal (accounting for up to 40% of cases in the Far East) ► diffuse
- It is unclear whether HCC arises from a regenerative nodule (via a dysplastic intermediate state) or as a de novo lesion

RADIOLOGICAL FEATURES

- *Larger lesions (>3cm):* these may contain fat ► they may demonstrate haemorrhage, thrombosis or necrosis
  - *Vascular invasion:* this can involve the portal vein (35%) or hepatic veins (15%)

US

- A hypo-, iso- or hyperechoic lesion in relation to the adjacent parenchyma (± a hypoechoic outer margin representing a fibrous capsule) ► larger lesions can be heterogeneous (due to any haemorrhage, necrosis or fat)
- *Colour Doppler:* internal high-velocity signals can be due to arterioportal shunting) ► a portal vein filling defect represents either thrombosis or intravascular tumour (arterial signals will only be demonstrated within tumour)

CT

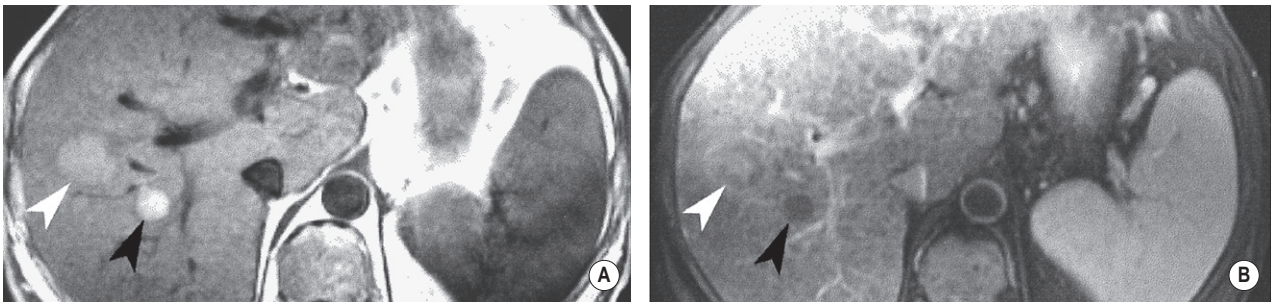
- *NECT:* ill-defined low attenuation lesions ► focal areas of internal calcification (7% of cases) ► there may be a hypoattenuating capsule
- *CECT:* enhancement is seen during the arterial phase, as it is a hypervascular tumour supplied via the hepatic artery ► it may demonstrate a mosaic enhancement pattern (with an enhancing grid-like pattern around a central lower area of attenuation) ► it will become hypoattenuating to the liver parenchyma during the portal phase
  - *Portal venous invasion:* arterioportal fistulae ► periportal streaks of high attenuation ► dilatation of the main portal vein (or its major branches) ► enhancement of any thrombus or detection of intrathrombus arterial flow
- *Lipoidal CT:* HCC foci will retain lipoidal (as there is no biliary drainage) and will be clearly seen when imaged 7–14 days later

MRI

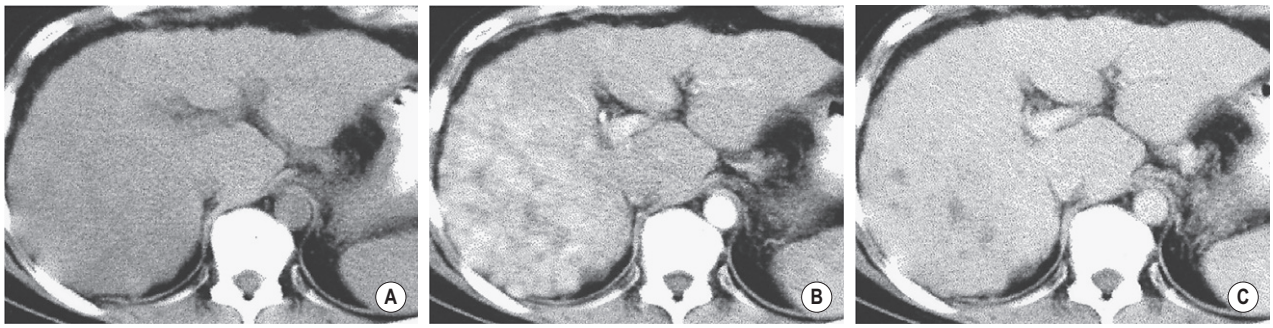
- T1WI: lesions less than 1.5cm are often isointense, whereas larger lesions may be hyperintense secondary to lipid, copper, or glycogen
  - Fatty metamorphosis in a cirrhotic nodule is suspicious for HCC
- T2WI: mild to moderate high SI (and possibly heterogeneous). Most lesions are hyper- or isointense
- T1WI + Gad: lesions <2cm in diameter can demonstrate homogeneous intense enhancement during the arterial phase, whereas larger lesions more often demonstrate heterogeneous enhancement
  - During the portal venous and equilibrium phases, a HCC will show rapid loss of enhancement (becoming iso- or hypointense relative to the liver) – this feature is very suggestive of malignancy ► venous washout is not displayed by regenerative and dysplastic nodules
  - *Atypical regenerative nodules*: these may cause confusion as they can also enhance during the arterial phase ► however they will be low SI on T2WI (due to iron accumulation – the so-called 'siderotic nodules')
    - The development of malignant foci within these nodules is suggested by the development of focal areas of high SI or heterogeneity within the low SI nodule
- DWI: variable appearance that depends on their histologic make-up
  - Well-differentiated tumors are often isointense
  - Moderately to poorly differentiated tumors are more often hyperintense
- **FDG PET:** this is relatively non-specific for HCC and is not widely used
- **DSA:**
  - This is used for preoperative assessment ► it defines the arterial and venous anatomy and evaluates any portal or caval involvement
  - HCC is usually a vascular lesion demonstrating dilated feeding arteries, abundant abnormal vessels or arteriovenous shunting
    - *Portal vein invasion:* a 'threads and streaks' appearance

PEARLS

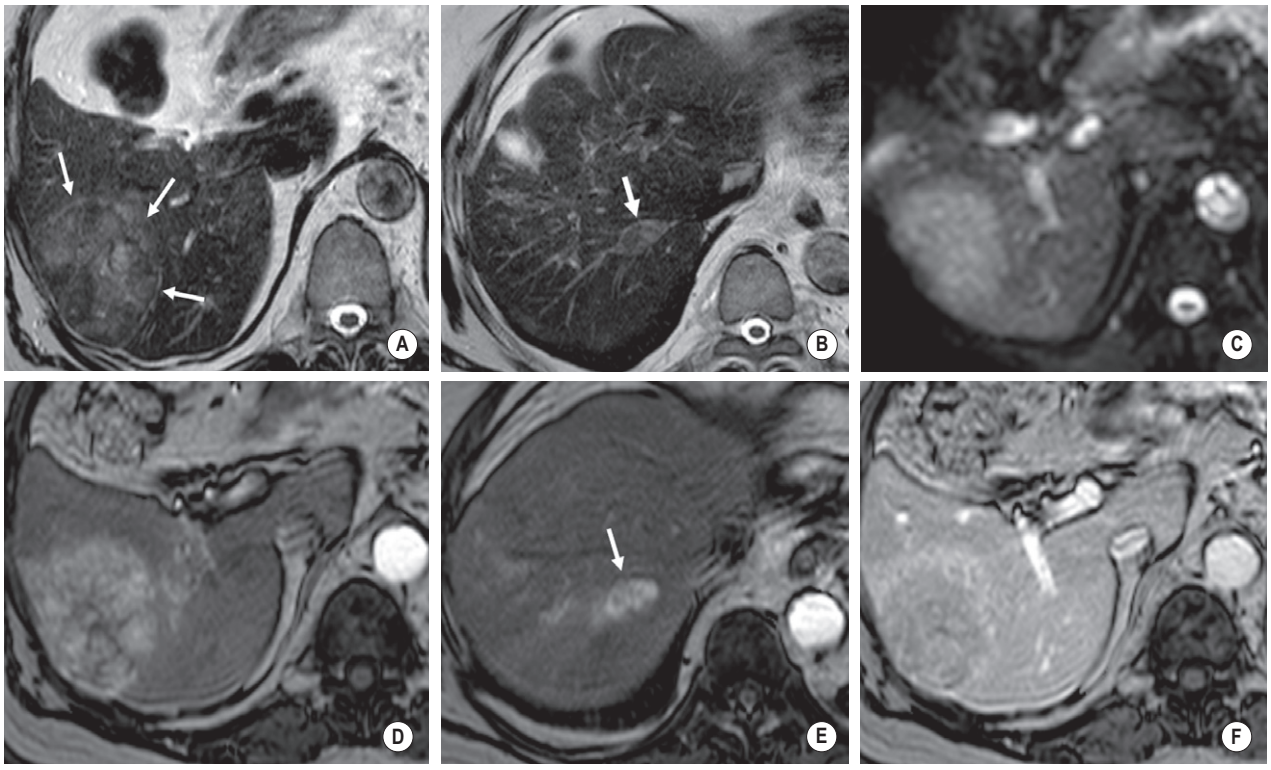
- The incidence parallels the prevalence of local predisposing conditions (in particular chronic hepatitis B and C)
- Serum α-fetoprotein (AFP) may or may not be elevated in HCC ► AFP may also be elevated with simple cirrhosis
- It commonly metastasizes to the lungs and bone



Hepatocellular carcinoma and regenerative nodule. T1WI (A) and T2WI (B) demonstrating an HCC (white arrowhead) and an adjacent atypical regenerative nodule (black arrowhead). Although the majority of hepatomas have decreased SI on T1WI, occasionally they have increased signal, thought to relate to fat or glycogen content. Note the heterogeneity in the hepatoma, particularly on T2WI.*



Multifocal HCC. A triphasic CT. NECT (A), arterial phase (B) and portal phase (C) examination demonstrates the multifocal and extensive nature of the tumour, which is only fully apparent during the transient enhancement of the arterial phase.*



HCC. (A) T2WI showing a heterogeneous high SI HCC (arrows). (B) T2WI. The right hepatic vein is expanded by high SI tumour extension (arrow). (C) DWI confirms restricted diffusion (also within the right hepatic vein - not shown). (D) Following IV gadolinium, there is heterogeneous enhancement. (E) The tumour within the right hepatic vein also enhances (arrow). (F) The HCC demonstrates washout on delayed imaging.

METASTASES

Definition

- The liver is a common site for metastases from many primary cancers (usually due to haematogenous spread)
  - *GI tract tumours:* these metastasize via the portal vein ► there is evidence for blood flow separation within the portal vein as right–sided colon cancers are more likely to spread to the right lobe (with left–sided tumours spreading to either the right or left lobes)
  - *Non-GI tract tumours:* these metastasize via the hepatic artery ► both lobes are equally affected
- Although a metastasis will derive its vascular supply from the hepatic artery it will usually be less vascular than the adjacent liver parenchyma

Radiological features

- Metastases can be difficult to radiologically detect and characterize if they measure less than 5mm in size (particularly in distinguishing them from a biliary hamartoma)
  - FDG PET does not improve the sensitivity (as there is a relatively high normal background liver uptake) but is useful in detecting extrahepatic metastases
- Metastases can demonstrate a wide range of appearances but they will usually demonstrate growth on serial imaging, multiplicity, and a variation in size

US Homogeneous or heterogeneous mass lesions ► they can be hyperechoic (mimicking a haemangioma) or hypoechoic (mimicking a simple cyst) ► central necrosis can cause a partly cystic appearance ► calcification can be seen in mucin-secreting metastases from the GI tract

- *'Target' appearance:* there may be a surrounding rim of reduced reflectivity

CT The majority of metastases are of low attenuation on unenhanced and portal phase imaging ► hypervascular tumours may show transient arterial enhancement, becoming isoattenuating to liver during the portal phase ► central necrosis, rim enhancement and calcification (in mucin-secreting metastases of GI origin) can also be demonstrated

- A <5mm low attenuation lesion within the liver is more likely to represent a simple cyst (unless a metastasis is purely cystic it is unlikely to be of low enough attenuation to be visible at such a small size)

MRI The signal intensity of a metastasis roughly parallels that of the spleen

- T1WI: hypervascular metastases are moderately hypointense ► haemorrhagic metastases can demonstrate hyperintensity
  - Perilesional fat deposition has been specifically described with hepatic metastases from a primary pancreatic insulinoma and is thought to be related to the effects of insulin
- T2WI: hypervascular metastases are usually markedly hyperintense and may be cystic or necrotic
- T1WI + Gad: similar enhancement characteristics as for CT
  - With paramagnetic iron oxide agents the normal liver parenchyma is of low SI (due to Kupffer cell uptake) – this will make a metastatic lesion more obvious
- DWI: hyperintense

Colloid scintigraphy There is reduced activity (metastases lack Kupffer cells)

Pearls

- *Cystic metastases:* ovarian tumours (the most common) ► carcinoma of the colon ► teratoma ► metastatic squamous tumours
- *Hypervascular metastases:* breast ► renal ► thyroid ► neuroendocrine tumours ► melanoma
- *Calcified metastases:* mucinous tumours of the GI tract ► endocrine pancreatic carcinoma ► osteosarcoma
- *Haemorrhagic metastases:* colon ► thyroid ► breast ► choriocarcinoma ► melanoma ► RCC
- After the initiation of chemotherapy, metastases can exhibit a less aggressive enhancement pattern that can mimic a haemangioma (including early peripheral nodular enhancement and delayed retention of contrast material)
  - A key distinguishing feature of chemotherapeutically treated metastases is an early, intact peripheral rim of enhancement (unlike the discontinuous peripheral enhancement seen with a haemangioma)
- Hypervascular metastases classically show marked T2 hyperintensity and restricted diffusion (compared with FNH and adenoma) ► they will wash out on delayed enhanced images (unlike a haemangioma)

ANGIOSARCOMA

Definition

- A rare malignant vascular hepatic neoplasm derived from the endothelial cells and which can form vascular derivatives, cavernous spaces, or solid masses
- It is associated with exposure to polyvinylchloride, arsenic and Thorotrast contrast medium

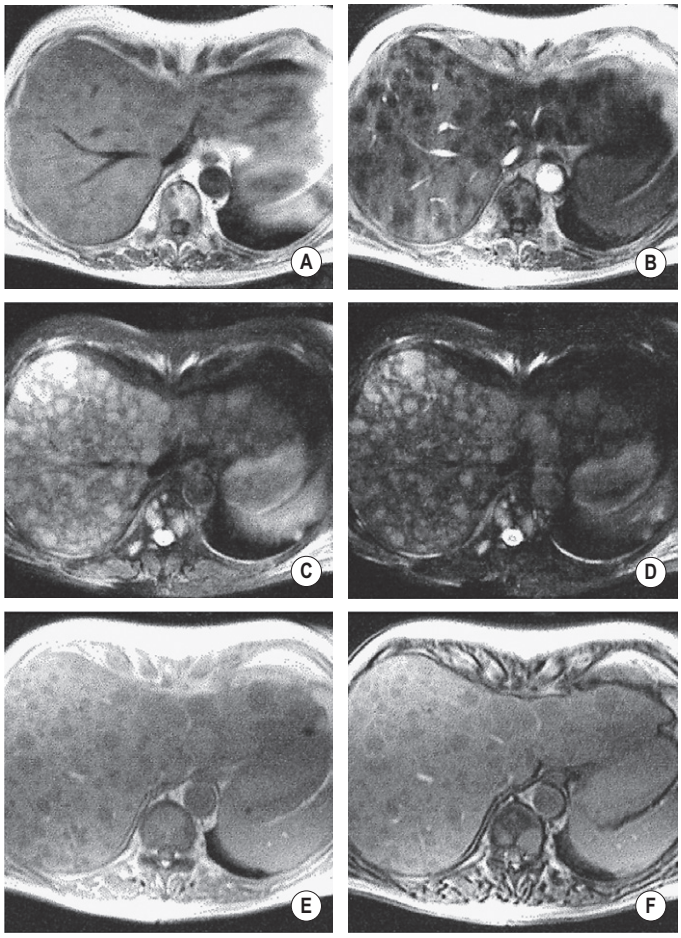
Radiological features

CT It can appear as an infiltrating mass demonstrating heterogeneous enhancement ► it can occasionally present in a diffuse form that is not easily detected with imaging

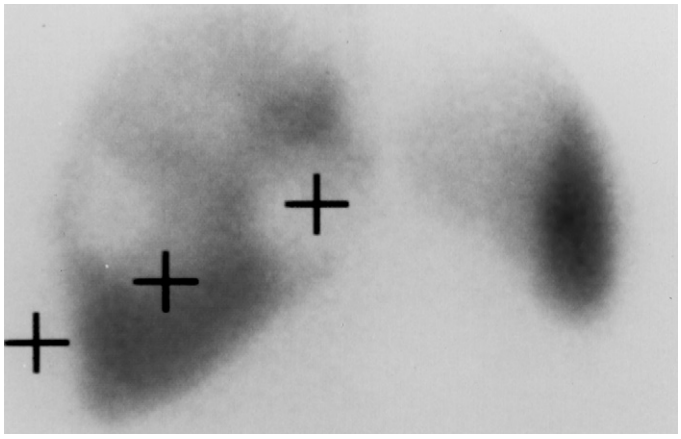
- Background thorotrast exposure causes heterogeneously increased attenuation within the liver, perihepatic lymph nodes and spleen

MRI It can present as a large mass or as multiple nodules

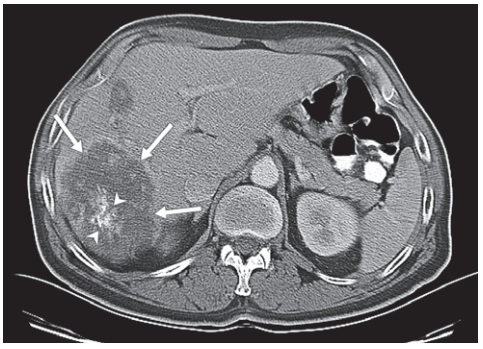
- T1WI: low SI ► T2WI: high SI ► T1WI + Gad: heterogeneous enhancement



Multiple metastases. MRI demonstrates lesions within both the liver and the vertebral bodies. They are of reduced SI on T1WI (A) gradient-echo images (B), moderately increased SI on T2WI images (C, D), and do not significantly change in relation to the splenic signal on in- (E) and out- (F) of-phase gradient-echo imaging, indicating the lack of any lipid content.

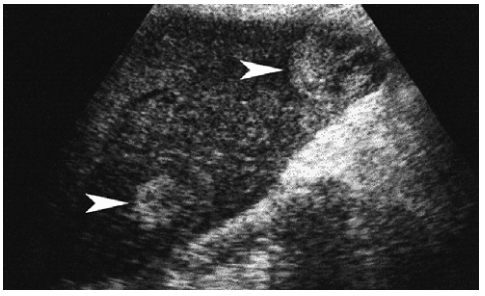


Multiple metastases. Anterior view demonstrates a significantly enlarged liver with multiple, large ('cannonball'), photopenic areas that are metastases from a colonic carcinoma. The overall liver uptake of sulphur colloid is decreased, resulting in colloid shift towards the spleen and, to a lesser extent, reticuloendothelial cells in bone.

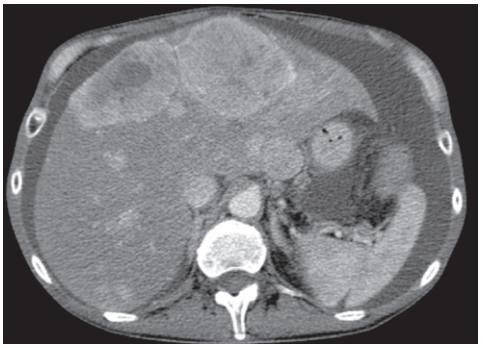


Hepatic metastatic lesion (arrows) from a mucinous adenocarcinoma of the rectum shows calcifications within the centre (arrowheads).••

Liver metastases. Portal phase CT study demonstrates multiple low attenuation lesions, most likely multiple metastases, in a patient with known colorectal malignancy.*



Echoreflective liver metastases. Metastatic carcinoid lesions of typical increased echoreflectivity (arrowheads). These lesions are usually of increased vascularity and can demonstrate arterial phase enhancement on CT and MRI.*



CECT of the liver demonstrating multiple hypervascular liver metastases. Ascites is also present.

FIBROLAMELLAR CARCINOMA (FLC)

Definition

- A hepatic tumour composed of sheets of fibrosis and numerous eosinophilic hepatocytes ► it arises spontaneously with no predisposing factors, and occurs within an otherwise normal liver (cf. HCC) ► there are no elevated AFP levels
  - It was previously classified as a variant of HCC – it is now considered a separate entity

Radiological features

- It is often a large, lobulated, well-defined tumour containing a central fibrous scar (with punctate calcification in >50% of cases) ► it is usually a solitary lesion

US Increased reflectivity (with a central scar of high reflectivity and a related acoustic corridor if calcification is present)

CT A low attenuation well-defined lesion with an even lower attenuation central scar (demonstrating radial components) ► there is punctate calcification of the central scar in ⅔ of cases (this is rare in FNH)

CECT There is moderate enhancement (± delayed scar enhancement)

MRI T1WI/T2WI: there is a low SI scar (cf. FNH with a high SI scar on T2WI)

Pearls

- There is a higher 5-year survival rate than seen with HCC (60% vs 30%) ► this is possibly due to a younger age at presentation and a lack of background liver disease

HEPATOBLASTOMA

Definition

- A hepatic tumour composed of primitive hepatocytes (often with mesenchymal components)
- It is the 3rd commonest childhood abdominal tumour (after neuroblastoma and Wilms tumour)

Clinical presentation

- Many are asymptomatic masses ► advanced tumours are associated with anorexia, weight loss, pallor, anaemia, and abdominal pain ► 20% of patients have metastases at presentation
- Patients are usually <3 years at presentation (M:F 2:1)

Radiological features

AXR Calcification is seen in 50% of cases

US A heterogeneous mass of mixed high and low reflectivity ► it may demonstrate calcification, cystic areas of necrosis, or a pseudocapsule ► the lesions can be small, large, single or multiple ► the tumours can splay or infiltrate the IVC, hepatic or portal veins

CT A mixed low attenuation lesion (± calcification) ► there can be peripheral rim enhancement

MRI T1WI: heterogeneous low SI (haemorrhage may demonstrate high SI) ► T2WI: high SI with hypointense fibrous septae

Scintigraphy ^{99m}Tc-sulphur colloid scintingraphy demonstrates activity during the angiographic phase and a photopenic area on delayed imaging

Pearls

- The tumour is usually associated with a markedly elevated serum AFP level (in over 75% of cases)
- **Associations:** Beckwith–Wiedemann syndrome (chromosome 11) ► familial adenomatous polyposis (chromosome 5)
- **Diagnosis:** percutaneous needle biopsy
- **Differential:** haemangioma ► metastatic neuroblastoma ► mesenchymal hamartoma ► hepatocellular carcinoma

EPITHELIOID HAEMANGIOENDOTHELIOMA

Definition

- A malignant tumour of vascular origin composed of 'epithelioid' endothelial cells
- It predominantly affects female adults and is associated with oral contraceptive pill use and vinyl chloride exposure
- It is not to be confused with an infantile haemangioendothelioma

Radiological features

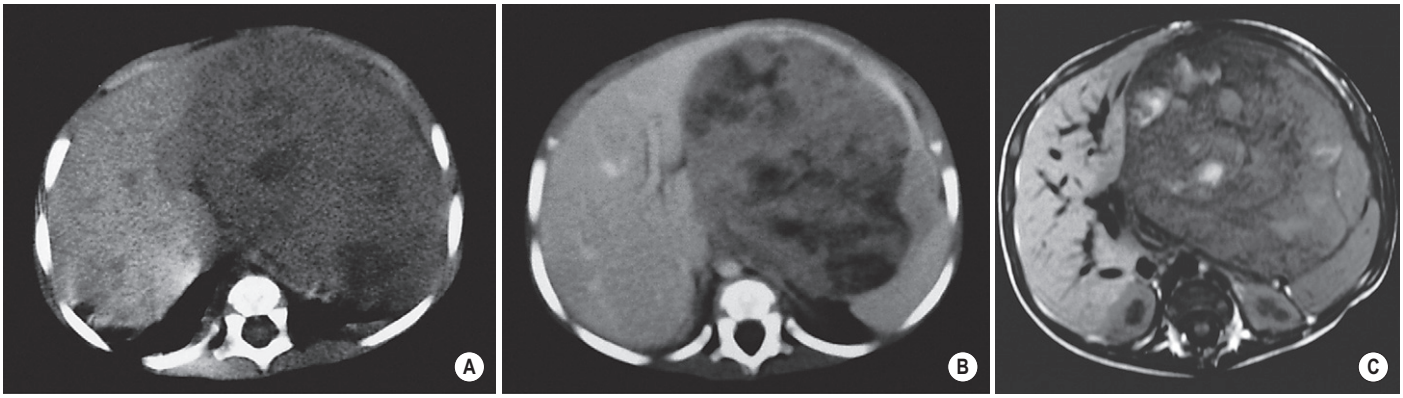
- The lesion appears as multiple peripherally situated nodules that may coalesce and cause capsular retraction, with compensatory hypertrophy of the uninvolved liver segments

US Solid hypoechoic lesions

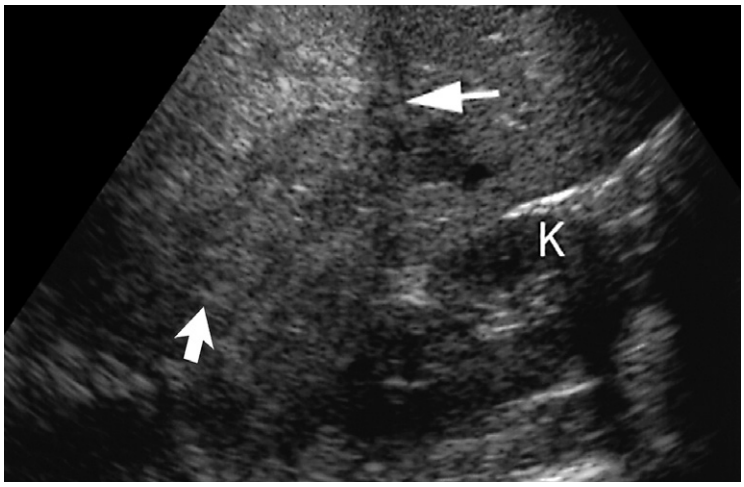
NECT Multiple low attenuation peripheral heterogeneous areas (± calcification)

CECT Nodular rim enhancement with a surrounding low attenuation 'halo'

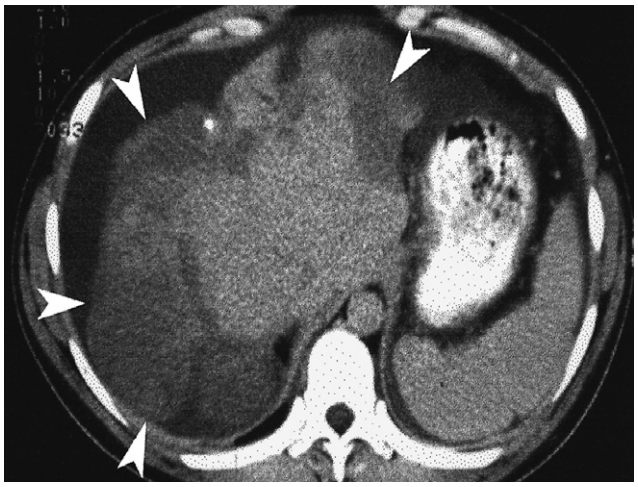
MRI T1WI: low SI ► T2WI: moderate high SI



Hepatoblastoma. (A, B) Large, low-density solid heterogeneous mass seen on CT without calcification and with patchy enhancement in a 17-month-old boy. (C) MRI. T1WI: low-SI large mass with areas of increased SI consistent with blood ► low-signal internal septae are also seen.*



Hepatoblastoma. Parasagittal view through the right lobe of the liver showing a solid echogenic mass (arrow) compressing the IVC (thick arrow). K = kidney.*



Epithelioid haemangioendothelioma. CT demonstrates peripheral low-attenuation lesions (arrowheads) that have coalesced to form a rind of tumour enclosing the central normal liver parenchyma. The patient presented with Budd–Chiari syndrome secondary to the tumour, diagnosed on needle biopsy and confirmed at subsequent liver transplantation.*

(A) A patient with fibrolamellar hepatocellular carcinoma showing a large low-density lesion with marginal enhancement on the arterial phase. (B) There is greater enhancement on the portal phase. (C) Near isodensity is seen on the venous phase.••

BUDD–CHIARI SYNDROME

DEFINITION

- A syndrome of global or segmental hepatic venous outflow obstruction which is secondary to obstruction of the IVC (usually by a membrane or thrombus), or by occlusion of the major hepatic vein branches (usually by thrombus)
  - *Type I:* occlusion of the IVC (± hepatic veins)
  - *Type II:* occlusion of the major hepatic veins (± IVC)
  - *Type III:* occlusion of the small centrilobar veins
- **Other causes:** congenital membranes or webs within the IVC (webs can also occur following a long-standing IVC thrombosis) ► oral contraceptive use or pregnancy ► coagulopathies (e.g. polycythaemia, thrombotic thrombocytopenic purpura, or sickle cell disease) ► tumour-induced hepatic vein compression ► hepatic vein trauma or surgery ► constrictive pericarditis ► right heart failure

CLINICAL PRESENTATION

- Acute hepatic vein obstruction can present with hepatomegaly, abdominal pain and ascites
- There can be a more insidious presentation with features of secondary portal hypertension and jaundice

RADIOLOGICAL FEATURES

Acute

US Hepatomegaly ► thrombus within the major veins (this can give an unequivocal diagnosis during the acute phase) ► abnormal collateral veins passing between the major hepatic veins ► poor visualization of the hepatic veins or of the flow within them

- A damped hepatic venous waveform (a non-specific sign)
- A continuous reversal of flow within a main hepatic vein
- An enlarged portal vein
- Gallbladder wall thickening

CT The caudate lobe is often preserved with a normal attenuation and enhancement pattern (it has not had time to enlarge) ► there may be hepatic vein thrombus (± collateral formation) ► the hepatic veins may be difficult to identify

- NECT: the enlarged, congested peripheral liver is of a lower attenuation than normal
- CECT: a 'flip flop' enhancement pattern:
  - *Early:* prominent central and weak peripheral liver enhancement
  - *Delayed:* washout of the central liver, with enhancement of the liver periphery

Chronic

CT Peripheral liver atrophy with compensatory hypertrophy of the caudate lobe (the caudate lobe usually drains via separate veins directly into the IVC and inferior to the normal hepatic venous confluence) ► secondary portal hypertension

Other investigations

MRI T1WI/T2WI: a heterogeneous and congested peripheral liver ► a normal or hypertrophied caudate lobe

- MRA: this can assess vascular patency and direction of flow

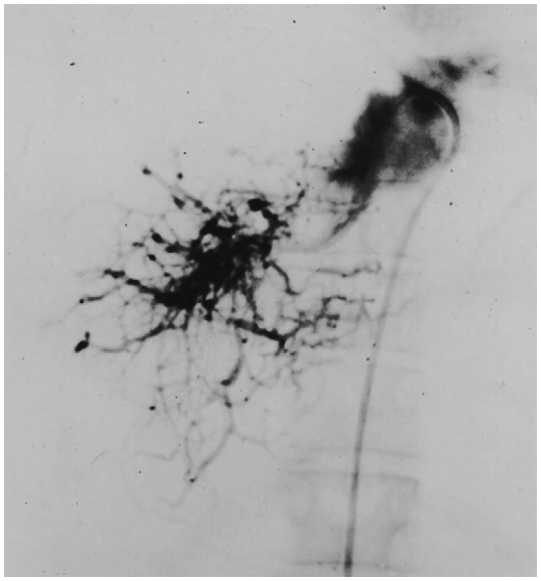
Sulphur colloid A normal or increased caudate lobe activity (there is reduced activity within the remainder of the liver) ► colloid shift to the spleen

DSA The venographic appearances are characteristic, resembling a 'spider's web'

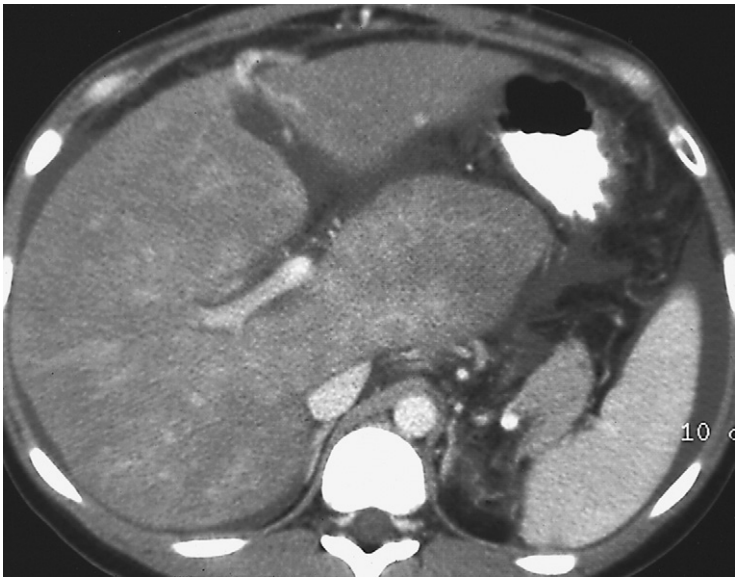
Cavography This can identify any IVC abnormality

PEARLS

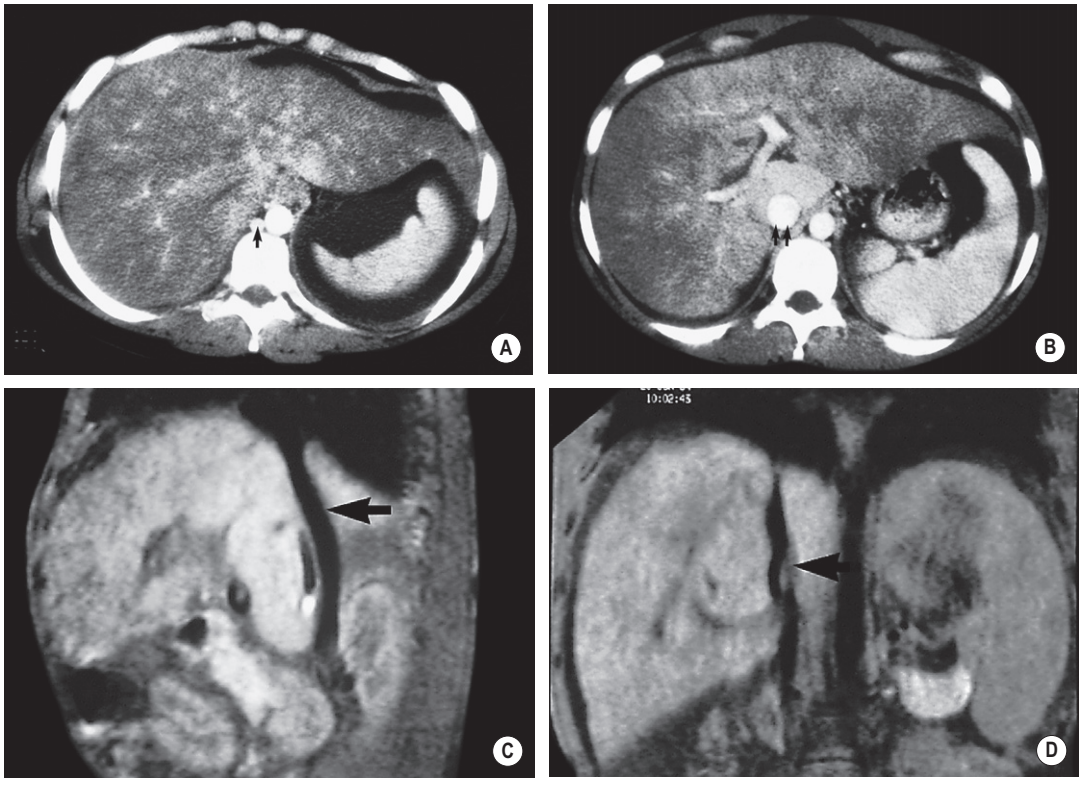
- Collateral venous channel development can allow some regeneration within the peripheral liver and caudate lobe, leading to variable findings (Budd–Chiari syndrome can be mistaken for extensive tumour involvement)
- The diagnosis in a patient with underlying cirrhosis is difficult – the related lobar and regenerative changes may distort the hepatic veins, making their visualization difficult
- A core needle biopsy is frequently required to exclude tumour and confirm the presence of central venous congestion and venous thrombi
- **Treatment:** liver transplantation ► some cases can be treated by interventional techniques (e.g. venous membranotomy, venous angioplasty and stenting)



Budd–Chiari syndrome. A catheter has been passed retrogradely into a right hepatic vein. Injection of contrast medium has outlined an extensive fine network of collateral vessels. This 'spider web' appearance is pathognomonic of the Budd–Chiari syndrome.*



CECT demonstrating Budd–Chiari syndrome. Note the preferential central enhancement of the liver, and the thrombus demonstrated within the IVC (arrow). A small amount of ascites is also present.



Budd–Chiari syndrome. CT and MRI. (A,B) CT shows patchy enhancement of the liver parenchyma, an absent inferior vena cava (IVC) at the level of the liver, with prominence of the azygos vein (A) (arrow) ► and more inferiorly enhancement of the caudate lobe with distension of the obstructed IVC (two small arrows) (B). (C,D) Compared with the previous patient, these sagittal (C) and coronal (D) MRI T1WI show a patent IVC (arrow) but absence of any recognizable hepatic veins in this child with more chronic features of Budd–Chiari syndrome.*

PORTAL VENOUS HYPERTENSION (PH)

DEFINITION

- A corrected sinusoidal pressure difference between the wedged (occluded) hepatic vein and IVC of >8mmHg (it is normally 4–8mmHg) ► causes can be defined as:
  - *Prehepatic:* portal vein thrombosis (this may cause PH or be the consequence of it)
  - *Hepatic:* cirrhosis
  - *Posthepatic:* Budd–Chiari syndrome ► congestive heart failure

RADIOLOGICAL FEATURES

US Ascites and distended mesenteric veins ► an oedematous gallbladder, stomach, and small bowel wall

- *Portal vein diameter:* this is >15mm (a normal diameter does not exclude the diagnosis)
- *Main portal vein mean peak velocity:* <10cm/s ► there is initially oscillating flow within the portal vein progressing to reversed (hepatofugal) flow
- *Splenomegaly:* this depends upon the degree of portosystemic shunting, and an absence of splenomegaly does not exclude the diagnosis
- *Portosystemic venous collaterals:* splenogastric ► gastro-oesophageal ► splenorenal ► a recanalized paraumbilical vein
  - *Recanalized paraumbilical vein:* unusual portal venous patterns may emerge due to the increased paraumbilical flow 'stealing' blood from the right portal vein (and resulting in hepatopetal right and hepatofugal left portal venous flow)

CT This is ideal for detecting the extrahepatic changes of portal venous hypertension, such as portosystemic shunts, and small bowel and gastric wall oedema

- Pre- and postcontrast images can assess the portal vein patency

MRI This is the non-invasive technique of choice if US is technically inadequate

- MRI and MRA: these can assess any GI tract changes as well as the hepatic and portal venous vasculature (± the presence of any shunt vessels)
- T1WI + Gad multiphase volumetric studies: this confirms the findings and also allows assessment of the flow direction (with breath-hold phase contrast or bolus tracking)

DSA This has been largely replaced by non-invasive techniques

PEARLS

- Imaging is often used to assess the patency of surgical shunts (e.g. between the splenic and left renal vein, and between the portal vein and IVC)
- A radiologically placed transjugular intrahepatic portosystemic stent shunt (TIPSS) is increasingly used for palliating portal hypertension

PORTAL VEIN THROMBOSIS

DEFINITION

- Thrombus formation within the portal vein can be idiopathic or due to: hepatic cirrhosis ► infection (portal pyaemia and acute cholecystitis) ► inflammation (pancreatitis and necrotizing colitis) ► tumour (HCC and pancreatic carcinoma) ► trauma ► coagulopathy ► surgery (liver transplantation)

CLINICAL PRESENTATION

- A patient can present with acute abdominal pain or with secondary complications (e.g. bowel infarction and ascites)
- There may be an occult presentation if there is already established cirrhosis (± portal hypertension and portosystemic shunt vessel formation)

RADIOLOGICAL FEATURES

Early An avascular solid lesion occluding and often expanding the portal vein

Late Contraction of the portal vein (which is often fibrotic or calcified)

- *Cavernous transformation:* multiple collateral vessel formation around the occluded portal vein
- Recanalization may make the discrimination between a tumour and pure thrombus difficult (a thrombosed portal vein or branch vein that remains enlarged is suspicious for tumor involvement)

US Acute thrombus is hypoechoic ► arterial signals within a thrombus is suggestive of tumour involvement (but may represent recanalization of the thrombus)

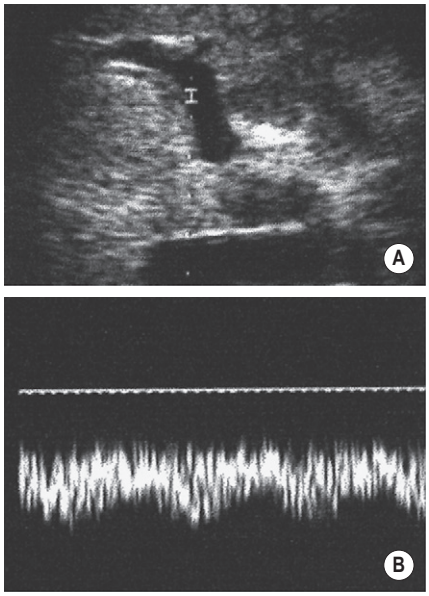
- Severe cirrhosis or fat infiltration may attenuate the acoustic beam to the extent that Doppler assessment is unreliable

MRI Time-of-flight and contrast-enhanced techniques can accurately demonstrate any portal vein thrombosis

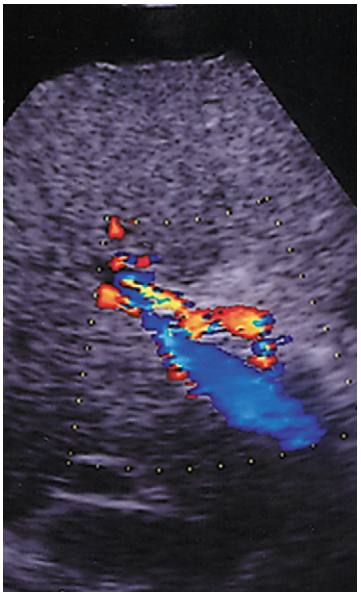
CT Unenhanced and portal phase imaging will visualize a portal vein thrombus and any underlying structural causes (e.g. tumour or pancreatitis)

PEARL

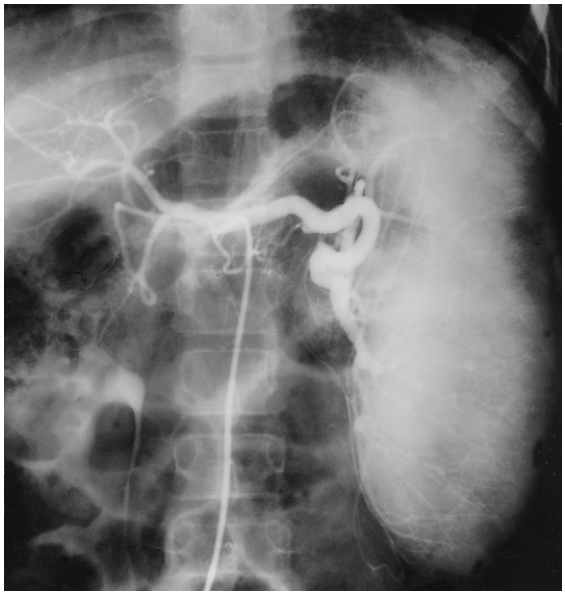
- Assessment of portal vein patency in a cirrhotic patient is important as it will influence the choice of surgical or radiological shunt procedure



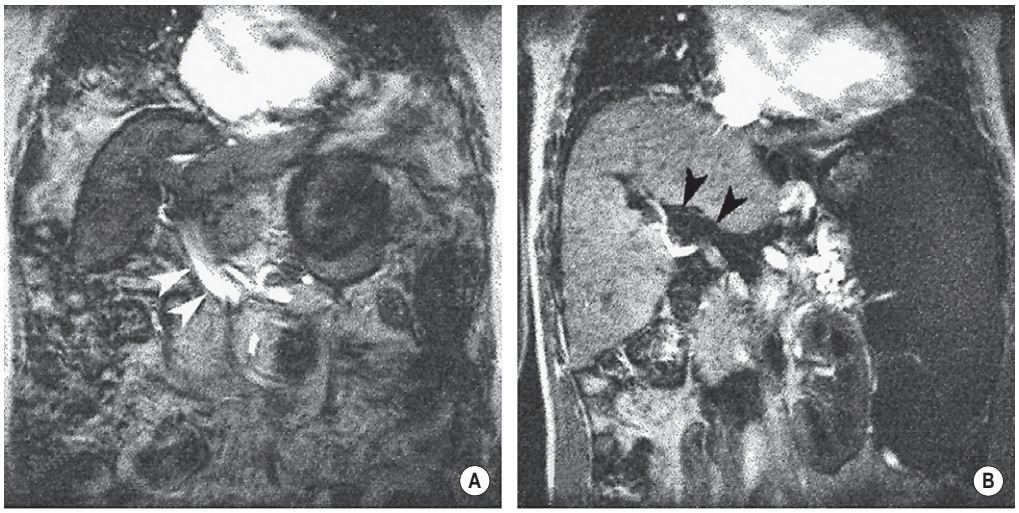
Portal venous hypertension: reversed portal vein flow. Duplex examination of the portal vein (A) demonstrates continuous reversed (hepatofugal) flow in the portal vein (B), usually reflecting underlying severe cirrhosis and portal venous hypertension with varices.*



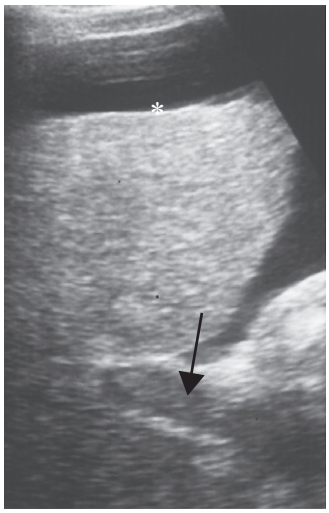
Note the irregular liver margin, coarse echoreflectivity and ascites in this cirrhotic liver, with normal forward flow (encoded red) within the hepatic artery and reversed flow (encoded blue) within the portal vein.



Coeliac angiogram in portal hypertension. Sparse liver arteries. Enlarged tortuous splenic artery with aneurysms on the main trunk and its divisions. Intrasplenic branches are stretched within a grossly enlarged spleen.†



Portal vein thrombosis. (A) Flow-sensitive coronal magnetization prepared gradient-echo sections in two patients with cirrhosis and ascites demonstrate a patent portal vein (white arrowheads). (B) Completely occluded portal vein (black arrowheads). MRA using either time-of-flight or contrast-enhanced techniques can be diagnostic when Doppler US is equivocal or technically limited.*



There is tumour thrombus (arrow) within the portal vein in this patient with multifocal hepatoma in a cirrhotic liver and ascites (asterisk).†

VENO-OCCLUSIVE DISEASE (VOD)

DEFINITION

- This results from obliteration of the central draining veins of the hepatic lobules by an inflammatory fibrotic process
- It usually occurs following chemotherapy for bone marrow transplantation (resulting in secondary portal hypertension)
  - Cirrhosis is uncommon in bone marrow transplant patients – therefore suspect the onset of VOD

RADIOLOGICAL FEATURES

- Imaging is used to exclude other causes of abnormal liver function
- It demonstrates non-specific features: hepatomegaly ► portal hypertension (the major hepatic veins are not usually involved)

PEARL

Diagnosis Biopsy (coagulation markers may be an effective alternative)

CECT showing hepatomegaly and ascites in a patient with veno-occlusive disease.

VASCULAR SHUNTS: ARTERIOPORTAL

DEFINITION

- A direct communication between branches of the hepatic artery and portal vein
  - Shunts can be misinterpreted as malignant lesions ► large shunts require embolization

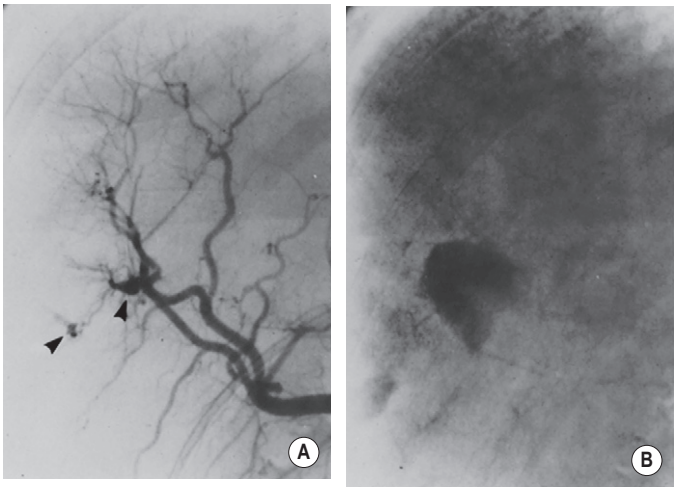
CAUSES

- A penetrating liver injury (e.g. percutaneous diagnostic and interventional procedures) ► cirrhosis ► portal hypertension ► tumours (e.g. a large HCC)

RADIOLOGICAL FEATURES

US An area of increased flow on colour Doppler imaging (± arterialization of the portal venous flow if the shunt is large enough) ► a lesion may appear rounded or wedge shaped

CECT/DSA An early enhancing focal lesion with early filling of the portal vein ► hepatic arterial blood entering a portal vein branch produces a cone-shaped segmental portal 'blush' within the surrounding parenchyma



Hepatic arteriogram in a patient who had undergone percutaneous cholangiography 2 days earlier. (A) Arterial phase: arrowheads point to small arterioportal fistulae. (B) Capillary phase: dense 'blushes' due to early portal venous staining.*

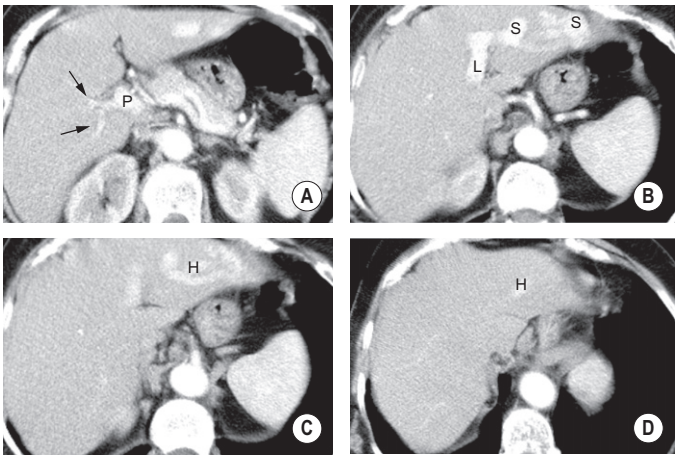
VASCULAR SHUNTS: INTRAHEPATIC PORTOSYSTEMIC

DEFINITION

- A direct communication between the branches of the portal venous system and systemic hepatic veins

RADIOLOGICAL FEATURES

- *Congenital cases:* multiple small portovenous shunts (1–2mm in diameter) within the periphery of an otherwise normal liver ► it may present with unexplained hepatic encephalopathy ► it is only detected with angiography
- *In association with portal hypertension and cirrhosis:* these are larger shunts typically between the right main portal vein and IVC ► their larger size allows detection with angiography, US and CECT



Intrahepatic portosystemic shunt. (A) The main portal vein (P) has fairly atrophic right branches (arrows). (B) This is because the left portal vein (L) is engorged as it directs the majority of flow through a portosystemic shunt (S). (C, D) The shunt directs flow into an early-filling left hepatic vein (H).

VASCULAR SHUNTS: ARTERIOVENOUS

DEFINITION

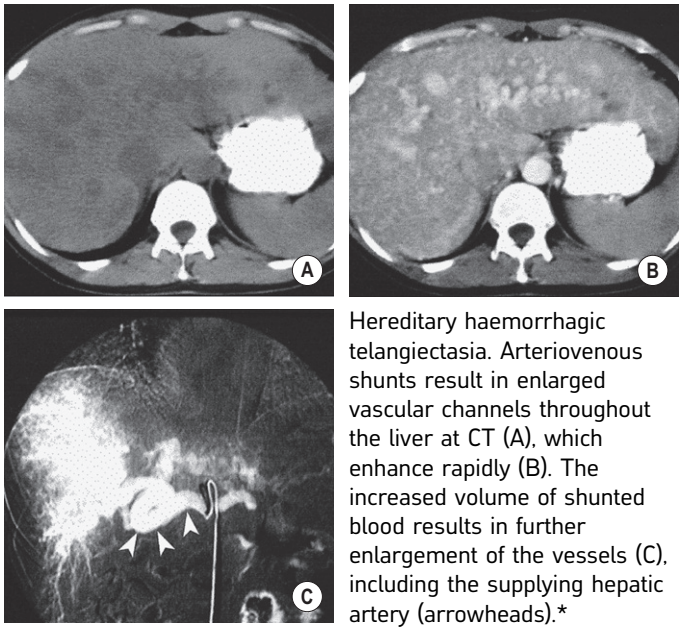
- A direct communication between arteries and veins without an intervening capillary bed
  - *Causes:* trauma ► tumours ► hereditary haemorrhagic telangiectasia (Osler–Weber–Rendu disease with multiple small intrahepatic arteriovenous shunts)

CLINICAL PRESENTATION

- They are often asymptomatic, but large shunts can lead to heart failure ► vascular dilatation can cause biliary obstruction and recurrent cholangitis ► ultimately hepatic necrosis may occur (and can be exacerbated by attempts at arterial embolization)

RADIOLOGICAL FEATURES

- Dilated hepatic arteries, hepatic and portal veins and with a tortuous vascular channel providing an intraparenchymal communication
  - *Small lesions:* these are only evident on DSA
  - *Larger lesions:* these are demonstrated with US (particularly Doppler studies), CECT and MRI



Hereditary haemorrhagic telangiectasia. Arteriovenous shunts result in enlarged vascular channels throughout the liver at CT (A), which enhance rapidly (B). The increased volume of shunted blood results in further enlargement of the vessels (C), including the supplying hepatic artery (arrowheads).*

HEPATIC TRAUMA

Definition Blunt or penetrating trauma may lead to an intraparenchymal laceration or haematoma, a subcapsular haematoma, or capsular rupture (with associated intraperitoneal haemorrhage)

Radiological findings

US This is more useful for follow-up rather than for diagnosis

- *Acute injury:* a parenchymal laceration with a related haematoma appears as an elliptical or irregularly shaped area of mixed low and high reflectivity (very recent haemorrhage may be relatively hyperechoic) ► free intraperitoneal fluid indicates a capsular rupture ► a subcapsular haematoma is well demonstrated

CT This is the investigation of choice ► it is able to assess the type of lesion and its anatomical relationship to the major hilar structures, the confluence of the hepatic veins and the IVC ► intraparenchymal lacerations and haematomas are again usually elliptical or linear in shape

- *NECT:* a low attenuation laceration ► high attenuation subcapsular and free intraperitoneal blood (recent haemorrhage is of higher attenuation than normal blood due to clot retraction)
- *CECT (arterial phase):* this is suggestive of a major vascular injury if a laceration involves the hilum or if there is a major perfusion deficit
- *CECT (portal phase):* this is mandatory to detect subtle lesions

Angiography This is only required when there is continuing haemorrhage (suggesting a major vessel laceration of a degree that is not immediately life-threatening) ► it can identify the source of any bleeding and permits embolization

MRI This is not routinely used ► it can demonstrate a parenchymal or subcapsular haematoma (especially when it is subacute as methaemoglobin increases the SI on T1WI) ► MRCP can assess the biliary system

Pearl

Unless the injury is life-threatening there is a trend towards conservative management

Complications Ischaemia and necrosis of the liver ► abscess formation ► haemobilia ► focal fibrosis ► calcification ► lobar or segmental atrophy

AAST liver injury grading system

Grade	Description
I	***Haematoma:*** subcapsular, <10% surface area Laceration: capsular tear, <1cm in parenchymal depth
II	***Haematoma:*** subcapsular, 10–50% surface area ► intraparenchymal, <10cm in diameter Laceration: 1–3cm in parenchymal depth, <10cm in length
III	***Haematoma:*** subcapsular, >50% surface area or expanding or ruptured subcapsular haematoma with active bleeding ► intraparenchymal, >10cm or expanding or ruptured Laceration: >3cm in parenchymal depth
IV	***Haematoma:*** ruptured intraparenchymal haematoma with active bleeding Laceration: parenchymal disruption involving 25–75% of a hepatic lobe or one to three Couinaud segments within a single lobe
V	***Laceration:*** parenchymal disruption involving >75% of a hepatic lobe or more than three Couinaud segments within a single lobe Vascular: juxtahepatic venous injuries (i.e. retrohepatic vena cava or central major hepatic veins)
VI	***Vascular:*** hepatic avulsion

LIVER ABSCESS

Definition

- A localized intrahepatic collection of pus
- It is usually secondary to portal pyaemia (e.g. pyogenic, fungal, or mycobacterial) ► immunocompromised patients are at an increased risk
- **Early:** It can mimic a solid tumour (e.g. metastases) ► it may require aspiration or biopsy for diagnosis
- **Late:** There is progressive central liquefaction with a surrounding inflammatory wall

Radiological features

US *Early:* a solid ill-defined lesion of low reflectivity ► *late:* there is a thickened irregular wall ► the necrotic centre generates sparse echoes

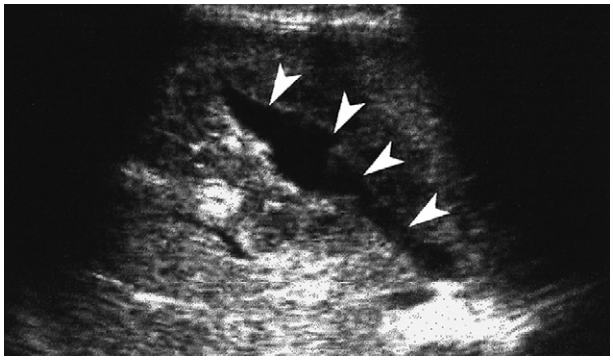
CT An ill-defined low attenuation lesion demonstrating rim enhancement (which may not be apparent once antibiotic treatment is started) ► when the central abscess liquefies it may be of water attenuation (and fail to enhance), appearing similar to a necrotic or cystic metastasis

MRI T1WI: low SI ► T2WI: high SI (often with a higher SI outer margin)

- With progressive liquefaction the central region will demonstrate increasingly low SI (T1WI) and high SI (T2WI)

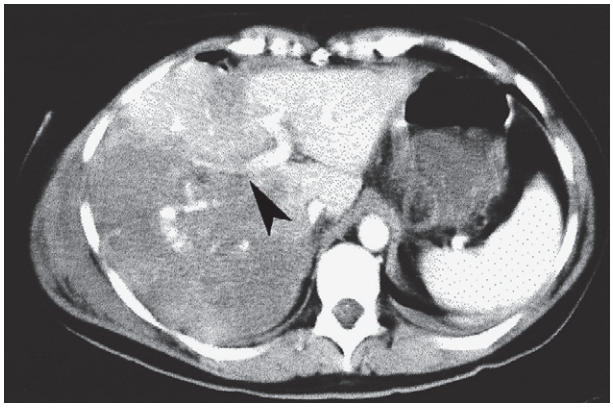
Pearl

Treatment Image-guided aspiration or drainage (+ medical therapy) ► surgery is rarely required

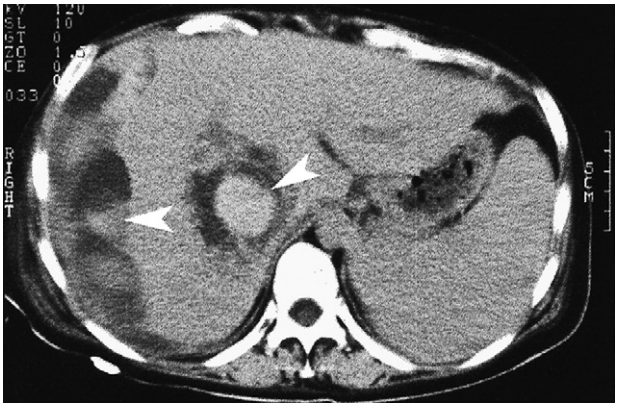


Intrahepatic laceration. Blunt hepatic trauma from a horse hoof has resulted in a linear laceration of the parenchyma clearly visible on US.*

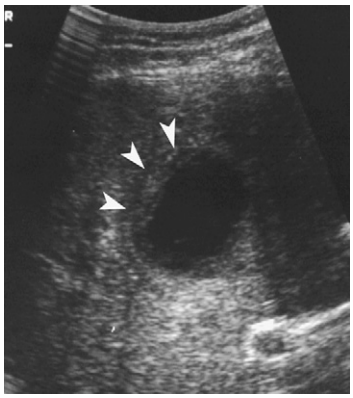
Subcapsular haematoma. T1WI in this patient is diagnostic for subacute haematoma (arrowheads) as methaemoglobin has produced increased SI within the organizing haematoma.*



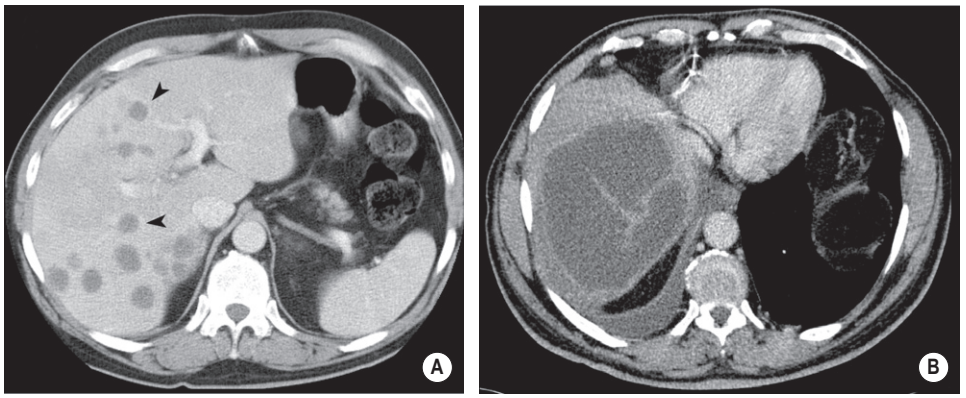
Portal vein trauma. This patient sustained liver trauma during a road traffic accident. Portal phase CT image demonstrates probable disruption of the right main portal vein (arrowhead) and failure of enhancement in the right lobe of the liver.*



Intrahepatic haematoma. Recent intraparenchymal haemorrhage following a liver biopsy is evident where sequestered blood has formed thrombi that undergo contraction, increasing globin density. This results in areas of subtly increased attenuation (arrowheads).*



Liver abscess. An abscess, with typically reduced echoreflectivity and a thickened irregular wall (arrowheads).



Liver abscess. Portal phase CT examinations in two different cases. (A) Multiple low attenuation lesions with ring enhancement (arrowheads) ► these appearances are often non-specific on CT and often overlap with those of metastatic deposits. (B) The presence of septae, central low attenuation, along with a sympathetic pleural effusion, aid the diagnosis.*

LIVER CYSTS

DEFINITION

- True hepatic cysts arise from abnormal development of the bile duct precursors (Meyenburg's complexes) which are lined by cuboidal epithelium
  - *Rare causes:* as a long-term sequelae of a parenchymal haematoma ► abscesses or if multiple as part of the spectrum of adult polycystic disease

CLINICAL PRESENTATION

- They are rarely symptomatic
- Large cysts may cause pain, become infected, or suffer internal haemorrhage

RADIOLOGICAL FEATURES

US

- **Simple cyst:** spherical anechoic structures with an imperceptible wall ► there is posterior acoustic enhancement ► there is no internal flow on Doppler settings
- **Complex cyst:** there can be internal echoes, thick septations, perceptible wall, or solid components ► this needs CT or MRI to characterize further
  - *Causes:* haemorrhagic cyst ► abscess ► cystic metastasis (e.g. ovarian) ► biliary cystadenoma (or cystadenocarcinoma) ► hydatid disease

CT A homogeneous (0–10HU) lesion with an imperceptible wall ► there is a lack of enhancement (internally or within the wall) ► there can be increased attenuation if it is a proteinaceous, infected, or haemorrhagic cyst

- Partial voluming may efface the characteristics of small lesions

MRI T1WI: low SI ► T2WI: very high SI (similar to CSF) ► T1WI + Gad: no enhancement

Scintigraphy Non-specific photopenic regions ► hepatobiliary iminodiacetic acid (HIDA) imaging may distinguish this from a choledochal cyst (which will show increased activity)

PEARL

Peliosis hepatis This is related to androgenic anabolic steroid use, and HIV with associated cutaneous bacillary angiomatosis (vascular proliferation containing bacteria) ► it is rare but increasing in frequency ► it affects the liver and other sites (e.g. the spleen)

- It is characterized by multiple small cystic lesions which demonstrate centrifugal or centripetal enhancement

HYDATID DISEASE

DEFINITION

- This follows liver infection with *Echinococcus granulosus* – a parasitic tapeworm transmitted to humans from dogs, sheep, foxes and other wild animals
- The larvae migrate from the gut and embed within the liver (and subsequently the lungs) where they encyst and develop, slowly provoking a surrounding inflammatory reaction ► they may remain occult for several years
  - *Endocyst:* the parasitic component – an inner germinative layer giving rise to the daughter vesicles
  - *Ectocyst:* the cyst membrane
  - *Pericyst:* the protective host fibrotic granulation tissue

RADIOLOGICAL FEATURES

XR Crescentic calcification within the pericyst ► complete calcification of all the cyst layers implies parasitic death

US The appearances can range from a simple cyst to a complicated cyst with any of the following features:

- *A heterogeneous mass:* the most common appearance
- *'Hydatid sand':* internal echogenic foci formed from dead dependent scolices
- *'Cyst with a cyst' appearance:* multiple daughter cysts
- *'Double rim' sign:* the pericyst and endocyst are seen as echogenic lines
- *Partial or complete detachment of the endocyst from the pericyst:* a floating membrane (partial) or the 'water lily' sign (complete)

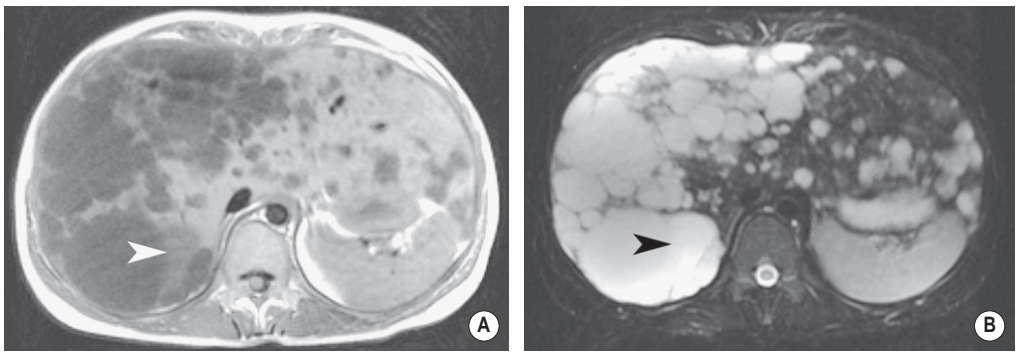
CT A well-demarcated low-density cyst of fluid attenuation ► there is enhancement of the cyst wall

MRI T1WI: low SI cyst contents with a low SI rim ► T2WI: high SI cyst contents with a low SI rim

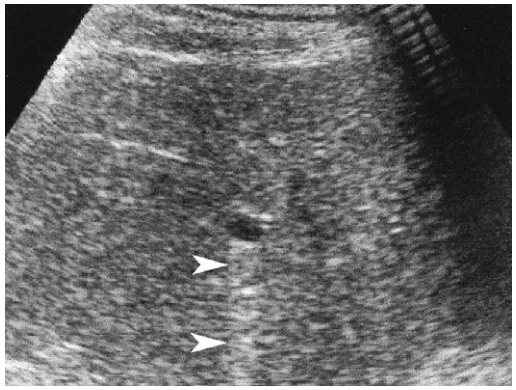
- MRI is insensitive to any calcification

PEARL

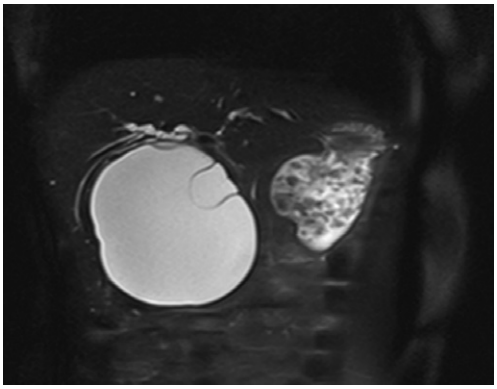
- Diagnosis is made with serological testing ► the risk of anaphylaxis with aspiration is less than previously thought



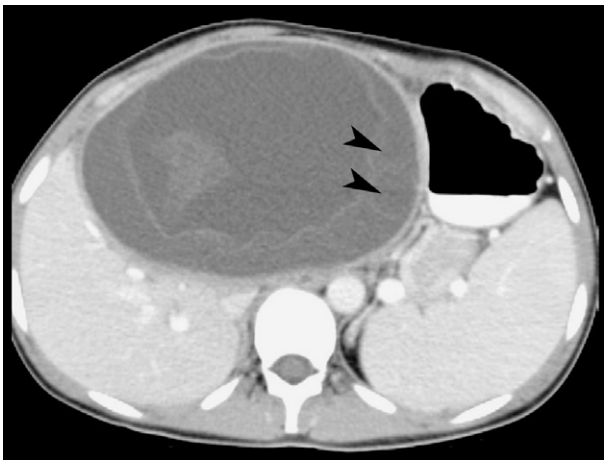
Polycystic liver disease. Multiple simple liver cysts are present and typically low signal on T1WI (A), and increased signal (greater than that of the spleen) on T2WI (B). Confusion may occur in the presence of haemorrhage, as this may increase the signal on T1WI (white arrowhead). In these circumstances the lack of enhancement following IV gadolinium DTPA may be diagnostic.*



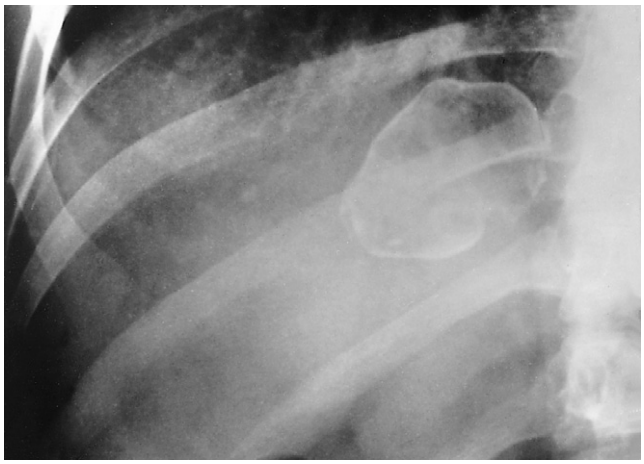
Simple liver cyst. On US simple cysts are well-defined areas of reduced echoreflectivity with no perceptible wall and posterior acoustic enhancement (arrowheads).



Biliary cystadenoma. Coronal T2 image demonstrating a large biliary cystadenoma with some simple septation seen medially.



Hydatid disease. Portal phase CT demonstrates a large cystic structure with a discrete wall, separated internal membranes and several 'daughter cysts' (arrowheads).*



Typical egg shell calcification of an hydatid cyst.*

HEPATIC CALCIFICATION

DEFINITION

- Focal benign parenchymal calcification is relatively common
- *Causes:* tuberculosis ► sarcoidosis ► pyogenic abscesses ► parenchymal haematoma ► giant haemangioma ► metastatic mucin-secreting adenocarcinomas (e.g. colonic) ► hepatoblastoma ► fibrolamellar hepatoma ► Pneumocystis infection (with widespread focal calcification)

RADIOLOGICAL FEATURES

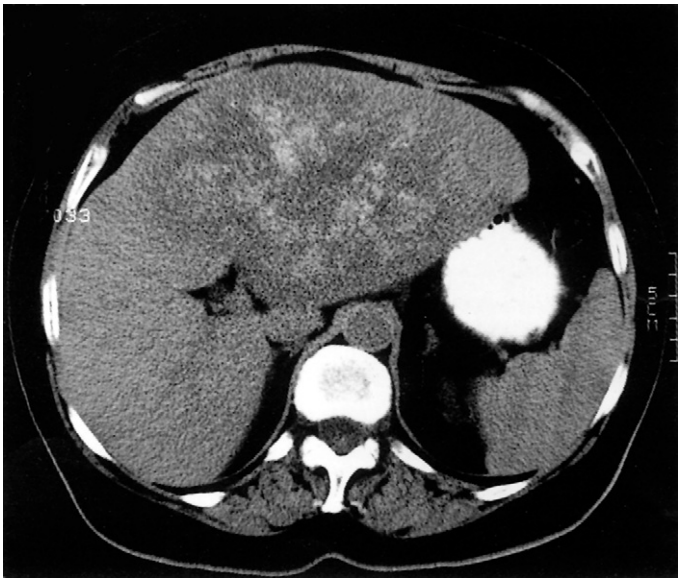
- Parenchymal calcification is usually well demarcated and surrounded by normal parenchyma

AXR Calcific densities

US Increased areas of reflectivity with a posterior acoustic shadow

NECT A high attenuation focus

MRI This is insensitive to calcification



Liver calcification. NECT showing a large metastatic deposit in the left lobe of the liver from a primary colonic adenocarcinoma. Faint calcification is visible in the metastasis, which could be masked following IV contrast medium enhancement.†

AEROBILIA

DEFINITION

- Gas present within the biliary tree
- *Causes:* a sphincterotomy ► a Roux loop procedure (allowing reflux of intestinal gas into the biliary tree)

RADIOLOGICAL FEATURES

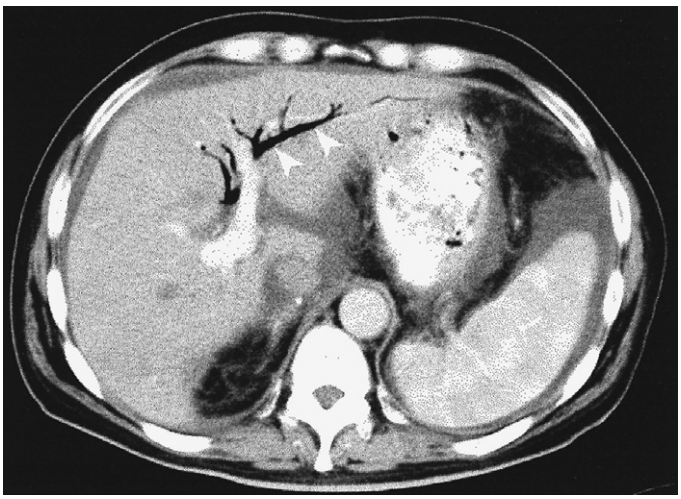
- A linear distribution of gas radiating from the hilum ► there is a gravity dependence with air predominantly located within the non-dependent parts of the biliary tree

US The biliary ducts are apparent as echogenic linear structures ► there is movement of any gas with respiration or patient position

CT This is extremely sensitive for the detection of air (which will measure −1000HU)

PEARL

- A rough approximation for small amounts of air (this is not valid for large amounts of air):
  - Biliary air tends to be more centrally located within the liver (due to centripetal biliary flow)
  - Portal venous gas tends to be more peripherally located within the liver (due to centrifugal portal venous flow)



Biliary duct gas. CT demonstrates clearly the low attenuation gas (arrowheads) in the non-dependent biliary tree.*

PORTAL VEIN GAS

DEFINITION

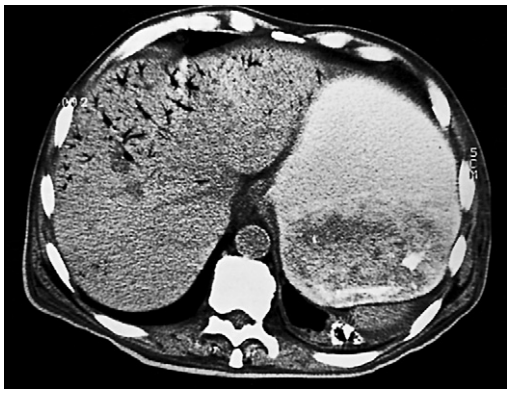
- Gas within the portal vein and its branches ► this arises when intestinal permeability increases together with an increase in the intestinal luminal pressure
- *Causes:* neonatal necrotizing enterocolitis ► gastric emphysema ► intestinal volvulus ► infection ► ischaemic bowel ► blunt abdominal trauma ► invasive abdominal malignancies ► duodenal perforation at ERCP ► colitis following a barium enema

RADIOLOGICAL FEATURES

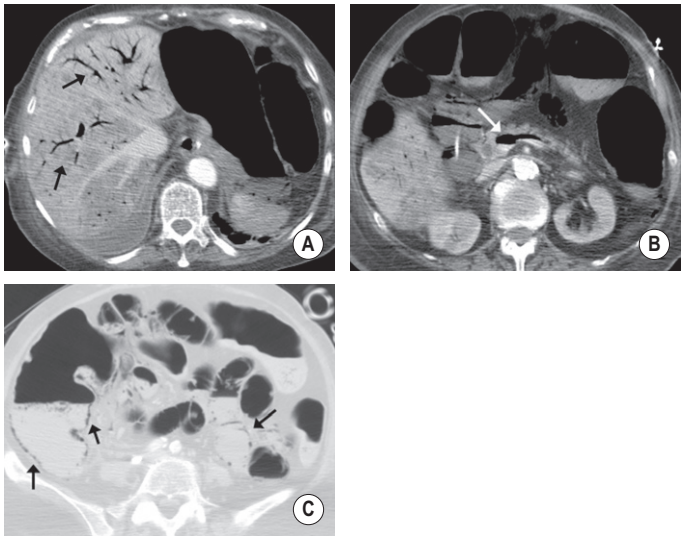
- Gas radiates out from the hilum ► there is less marked gravity dependence than seen with aerobilia

US This is the most sensitive modality and can demonstrate moving gas bubbles ► there is a high-pitched random bubbling and squeaking sound with focal alias artefacts seen on spectral display (as the gas bubbles overload the system receivers)

XR/CT This can detect portal vein gas if large amounts are present ► air (1000HU) is seen within the main portal vein and its branches



CT demonstrates air within the portal venous system of the liver in a postoperative patient unrelated to bowel necrosis.



(A) Axial CECT showing extensive portal venous gas (arrows) within the liver in a patient with ischaemic bowel. (B) CECT in same patient showing gas within the main portal vein (arrow) and splenic vein. (C) Axial CECT on lung windows showing extensive pneumatosis in the bowel wall in the same patient.

PARENCHYMAL GAS

DEFINITION

- Intrahepatic parenchymal gas
- *Causes:* gas-forming organism within an abscess or infarct ► post-traumatic ► hepatic arterial thrombosis following liver transplantation ► following embolization or thermal ablation of liver tumours

RADIOLOGICAL FEATURES

US An echogenic area with posterior acoustic shadowing ► it may be difficult to define its extent when large ► it can be confused with adjacent bowel

CT This is the most sensitive for its delineation

Large gas forming abscess within the right lobe of the liver demonstrating a clear air-fluid level. A small amount of ascites and a small pleural effusion are also present.

ORTHOTOPIC LIVER TRANSPLANTATION

- This is an established treatment for end-stage liver disease (with ≥90% 1-year survival and ≥80% 5-year survival)
  - ***Causes:*** cirrhosis secondary to infective hepatitis (the most common cause) ► autoimmune disease ► alcohol abuse ► Alagille's syndrome
- The majority of donor livers are of cadaveric origin ► however, demand exceeds supply, leading to the development of:
  - *Split-graft procedures:* a single organ donation to benefit 2 or more patients ► typically the left lateral segment for a child and the right lobe for an adult recipient
  - *Living donors:* donation of the full right lobe (most common) ► full left lobe ► left lateral segment
  - *Auxiliary transplantation:* 'piggy-backing' a graft alongside the native liver as a temporary measure (e.g. in reversible liver failure)

Technique

- A 'piggy back technique' is now the standard procedure, preserving the retrohepatic IVC and anastomosing the donor IVC patch to the recipient hepatic veins (which are formed into a common cuff)
- The hepatectomy plane is 1cm to either side of the middle hepatic vein and parallel to the 'principal plane'
- *Left lateral segment transplantation (paediatric):* donor segments II and III along with the left hepatic vein, left portal vein, left hepatic artery, and left bile duct are removed ► the middle hepatic vein and middle hepatic artery (segment IV) are preserved in the donor
- *Right lobe transplantation (adult):* entire donor right lobe, right hepatic vein, right portal vein, right hepatic artery, and right bile duct ► the middle hepatic artery (segment IV artery) and middle hepatic vein are preserved in the donor

Recipient assessment

- *Identify and characterize focal liver lesions:*
  - The Milan criteria is used for patients with cirrhosis and an increased risk of HCC: transplantation is performed if there is a single lesion (≤5cm) or 3 lesions (≤3cm)
  - A minimum of 2 different imaging techniques are required to identify a HCC (as this can be difficult to demonstrate within a cirrhotic liver) ► a biopsy is usually avoided due to the risk of tumour 'seeding'
  - Transplantation is rarely performed for a known cholangiocarcinoma (due to the poor prognosis)
- *Assess the patency of the portal vein and IVC:* this is usually with multiphase MRA or CTA
  - *Portal vein:* if this is occluded it is important to ascertain if the confluence of the SMV and splenic vein is involved as it will affect the surgical approach (vascular reconstructions can be used to 'jump' to this confluence and allow successful grafting despite an occluded main portal vein)
  - *IVC:* the extent of the involvement is important with Budd–Chiari syndrome
- *Identify anatomical variants*: MRA and CTA can be used to assess variant hepatic arterial anatomy (variant portal vein anatomy is less of an issue)

Living donor assessment

- Usually the left lobe is resected ► increasingly the right lobe segments are removed (but there is a greater mortality)
- Cross-sectional and three-dimensional (3D) imaging techniques are used to detect variants of the arterial, portal venous, hepatic venous and biliary systems
  - Anomalies that cross the planned surgical division plane are the most important and may lead to complications for both the donor and recipient

Perioperative imaging

- *The detection of early complications:* haemorrhage ► haematoma and abscess formation ► anastomotic breakdown
  - It also allows image-guided drainage and aspiration
- *Surveillance imaging:* this is performed regularly for high-risk groups (e.g. paediatric transplants and complex vascular reconstructions) ► it detects sudden complications that may respond to an immediate intervention (e.g. hepatic arterial occlusion)

Graft failure

- **Causes of early graft failure:**
  - *Primary non-function:* hepatocyte function fails to recover in the newly perfused graft despite vascular patency and good perfusion at the time of surgery ► it is influenced by hepatic steatosis
  - *Hepatic artery thrombosis:* this occurs in 3–5% adults and 5–15% of children ► Doppler US is the initial mainstay of diagnosis ► it presents with:
    - Catastrophic liver failure with infarction and abscess formation ► Biliary complications (e.g. a leak or stricture formation) ► Silently with no obvious sequelae
  - *Portal vein thrombosis and IVC occlusions:* these are relatively rare
  - *Acute rejection:* this is infrequent due to improved immunosuppression
  - *Overwhelming sepsis*
- **Causes of late graft failure:**
  - ***Causes:*** chronic rejection ► chronic ischaemia ► biliary anastomotic failure ► diffuse biliary disease due to sepsis ► recurrence of the underlying disease (e.g. primary sclerosing cholangitis or hepatitis C infection)
  - The diagnosis is often made on biopsy

INTERVENTIONAL LIVER RADIOLOGY

LIVER BIOPSY

- Spring-powered cutting sheath biopsy devices (e.g. BioPince™) collect more consistent core biopsies with less crush artefact than a manually operated system
- The traditional route for liver intervention uses either a horizontal right lateral intercostal approach, or an anterior subcostal approach (this does not traverse the pleura and is less likely to cause pulmonary complications)
- It is preferable to biopsy lesions using a route through intervening normal liver as this reduces the risk of haemorrhage
- Ascites surrounding the liver is a contraindication to biopsy
- US is usually used as the real-time capability allows faster positioning of the needle or catheter and allows selection of an oblique approach (CT is limited to axial imaging)
- ***Complications:*** haemorrhage ► pneumothorax ► biliary peritonitis ► perforation of the bowel or gallbladder ► haemobilia ► arterioportal shunt formation

PERCUTANEOUS TREATMENT OF LIVER METASTASES

- Percutaneous local ablation techniques:
  - *Injectables:* ethanol can be used to treat an unresectable HCC
  - *Lesional heating:* radiofrequency, microwave and interstitial laser photocoagulation cause tumour necrosis by inducing local hyperthermia and are used to treat both HCC and hepatic metastases ► radiofrequency electrodes and laser fibres can be inserted into a tumour under US, CT, or MRI guidance
  - *Freezing* (cryotherapy): this is less widely used
- **Follow-up:** imaging will determine if complete necrosis has been achieved (FDG PET may help to determine if there is any remaining tumour) ► tumour marker monitoring can be used if these were elevated preoperatively (e.g. serial α-fetoprotein and CEA for HCC and metastases, respectively)

VASULAR INTERVENTIONAL TECHNIQUES

Hepatic arterial embolization

- **Control of haemorrhage:** arterial bleeding may occur into the biliary tree, hepatic parenchyma, or peritoneal space
  - ***Causes:*** accidental or iatrogenic trauma ► neoplastic disease ► arteritides (e.g. polyarteritis nodosa)
  - Surgical ligation of the main hepatic artery may be insufficient (there may be an extensive collateral arterial supply)
  - It requires the selective catheterization and occlusion of the abnormal vessel ► embolization is usually performed with metallic coils
- **Tumour embolization:** primary or secondary tumours usually derive most of their blood supply from the hepatic arterial branches (the portal vein provides >70% of the parenchymal blood supply) – a tumour can therefore be rendered ischaemic by occluding its arterial supply ► this can provide good palliation in HCC and metastatic neuroendocrine tumours

Hepatic arterial infusion techniques

- *Transarterial embolization (TAE):* tumour blood supply is blocked using coils or microspheres placed via a selective hepatic arterial branch
- *Transarterial chemo-embolization (TACE):* as above, but alternating aliquots of a chemotherapy dose and embolic particles are selectively applied
- *Drug eluting beads (DEB):* these can deliver chemotherapy intra-arterially and block the blood vessels at the same time

Left or right portal vein branch embolization

- This induces lobar atrophy in the embolized pathological liver segments (e.g. right lobe) and hypertrophy in the remaining liver (e.g. left lobe) ► the volume of hypertrophied normal liver post-resection will now be sufficient to support life
- The non-embolized segments need to form at least 20–30% of the total liver volume pre-procedure to be able to undergo sufficient hypertrophy ► this value increases with background liver disease (e.g. cirrhosis)

Transjugular intrahepatic portosystemic stent shunt (TIPSS) insertion

- This involves the creation of a track between the portal vein and one of the hepatic veins (usually right), followed by insertion of an expandable stent to maintain its patency
- Its major indication is in patients with acute variceal haemorrhage which is resistant to emergency endoscopic sclerotherapy
  - *It can also treat:* severe abdominal ascites due to liver disease that is resistant to medical therapy ► Budd–Chiari syndrome ► hepatorenal syndrome
- ***Contraindications:*** encephalopathy ► right heart failure

TIPS Doppler indices	
Normal	***Abnormal***
Pulsatile turbulent stent flow	Non-pulsatile flow (<50cm/s) or absent stent flow
Mean velocity of 100–200cm/s	Localized high velocity (>220cm/s) with post-stenotic turbulent flow
Similar velocity at the portal and hepatic ends	Change in velocity of >100cm/s across the stent

DIFFUSION WEIGHTED MR IMAGING OF THE LIVER

Technique

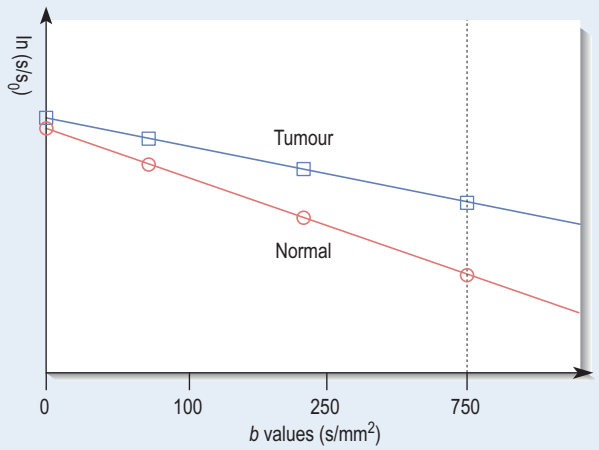
- DW MR imaging of the liver is usually performed prior to contrast material administration
- It uses a standard T2-weighted imaging sequence with application of a symmetric pair of diffusion sensitizing gradients on either side of the 180° refocusing pulse ► the use of parallel imaging techniques permits rapid imaging and reduction in motion artefact
- Although a number of imaging sequences can be applied, a single-shot spin-echo (SE) echo-planar technique is the most frequently used in combination with fat suppression (to reduce ghosting from respiratory motion and chemical shift artefact)
- Imaging may be performed during a breath hold, which attempts to freeze motion, or during free breathing with multiple signal acquisitions to reduce the effects of motion
  - *Breath-hold single-shot SE echo-planar imaging:* this is quick to perform
    - The whole liver can be evaluated in generally 1–2 breath holds of 20–30 s
    - *Disadvantages:* poorer signal-to-noise ratio (SNR) ► a greater sensitivity to distortion and ghosting artefacts ► a lower spatial resolution (with wider section thickness of 8–10mm) ► a limitation on the number of *b* values that can be included in the measurement
  - *Free breathing:* this can also be combined with respiratory ± cardiac triggering ► high-quality diffusion images can be obtained as cyclical respiration is a coherent motion
    - The liver is typically evaluated in 3–6 min ► multiple signal acquisitions results in an improved SNR ► therefore thinner image sections can be obtained and more *b* values accommodated
    - *Disadvantages:* slight image blurring ► volume averaging with a longer measurement time may impair assessment of lesion heterogeneity ► respiratory triggering increases the acquisition time as the images are only acquired during part of the respiratory cycle (increasing the risk of patient movement)
- **The major limitations of diffusion-weighted MR imaging:** a low signal-to-noise ratio (SNR) inherent in the technique ► a susceptibility to motion artefact
  - As single-shot SE echo-planar sequences are intrinsically sensitized to the motion of diffusion, they are also highly sensitive to other kinds of motion (e.g. respiration) ► in the left lobe of the liver, cardiac motion results in spin dephasing, leading to artefacts

Theory

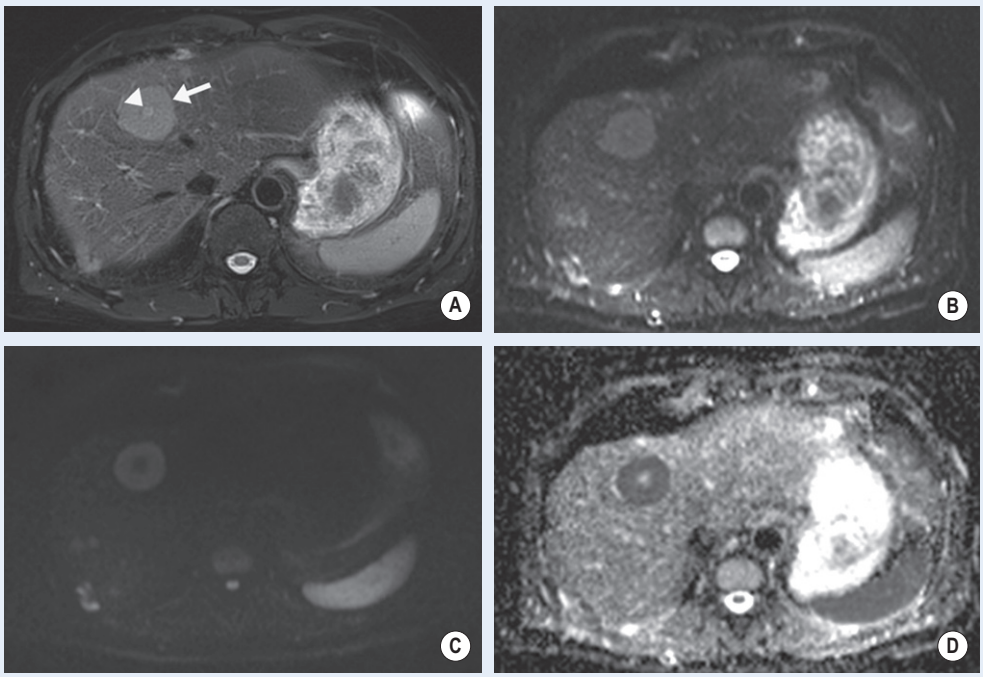
- **Static water protons:** these acquire an initial phase shift (the size of which is dependent upon the location along the sensitizing gradient) from the first diffusion-sensitizing gradient ► the second gradient will exactly reverse this phase shift as all protons remain in their original location
- **Moving water protons:** these acquire an initial phase shift (the size of which is dependent on the location along the sensitizing gradient) from the first diffusion-sensitizing gradient ► this is not exactly rephased by the second gradient (as the protons have moved from their original location and the second gradient is no longer a perfect match) ► this new reduced phase coherence results in attenuation of the measured signal intensity
  - Therefore the presence of water diffusion is observed as signal loss on DW MRI
- ***b* value:** this refers to the strength of the diffusion-sensitizing gradient, and is proportional to the gradient amplitude, the duration of the applied gradient, and the time interval between paired gradients ► the sensitivity of the diffusion sequence is adjusted by varying the *b* value (most readily achieved by altering the gradient amplitude)
  - *Small b values* (50–100 s/mm^2): this will result in signal loss in highly mobile water molecules ► the water molecules will have moved over relatively large distances by the time the rephrasing gradient is applied ► consequently they will not regain their original phase information after application of the rephrasing gradient.
    - The resulting images are referred to as 'black-blood' images due to the signal loss in the fast-flowing blood within vessels
  - *Higher b values* ($\geq$ 200 s/mm^2): as water movement in highly cellular tissues is restricted, such tissues retain their signal until higher *b* values are used
  - *Therefore applying a small diffusion-weighted gradient nulls the intrahepatic vascular signal (creating the so-called black-blood images) and improves the detection of focal liver lesions ► higher b values give diffusion information that helps focal liver lesion characterization*

- **Acquired diffusion coefficient (ADC):** for an individual voxel the ADC represents the slope (gradient) of a line that is produced when the logarithm of relative signal intensity of tissue is plotted along the y-axis versus *b* values along the x-axis ► the application of a greater number of *b* values will improve the accuracy of the calculated ADC
  - Each voxel will have an ADC value that can be combined visually as an ADC 'map'
- **T2 shine-through:** the signal intensity on diffusion-weighted images is dependent on water molecule diffusion and T2 relaxation time (it is based on a T2 sequence) ► therefore lesions with a high fluid content (e.g. cysts) can demonstrate high signal intensity even at high *b* values
- **DW MRI and tumour treatment:** effective tumour treatment results in an increase in the ADC value ► a transient reduction in ADC within 24–48 h after initiation of treatment has been observed (due to acute cell swelling) ► the ADC will subsequently decrease (due to tumour repopulation, fibrosis or tissue remodelling)
- **DW MRI imaging is a marker of cellularity,** therefore:
  - Benign solid lesions (e.g. FNH) may display restricted diffusion
  - Necrotic malignant lesions can demonstrate high ADC values
  - Therefore, DW MRI is most effectively interpreted in conjunction with other conventional MRI sequences.

b value	ADC	Cause
High	Low	Cellular tissue or tumour
Low	High	Cystic or necrotic tissue
High	High	T2 shine-through
Low	Low	Artefact or fat



A graph showing the relationship of signal intensities (y-axis) vs *b* values (x-axis). The gradient of the line represents the ADC for a tissue, and the gradient of the line is steeper for normal tissue than tumour.



Diffusion imaging of a liver metastasis. (A) T2 fat saturated image showing a high signal metastatic deposit (arrow) with a necrotic fluid filled centre (arrowhead). (B) At low *b* values, the metastasis is of high signal, demonstrating restricted diffusion, and the necrotic centre is also of high signal due to 'T2-shine through'. (C) At higher *b* values, the hypercellular periphery is of slightly reduced signal intensity (but still higher than the adjacent normal tissue). The necrotic centre loses proportionately more signal. (D) The hypercellular (restricted diffusion) nature of the periphery is confirmed by the signal loss on the ADC map, and the central high signal confirms the non restricted fluid nature of the necrotic centre.

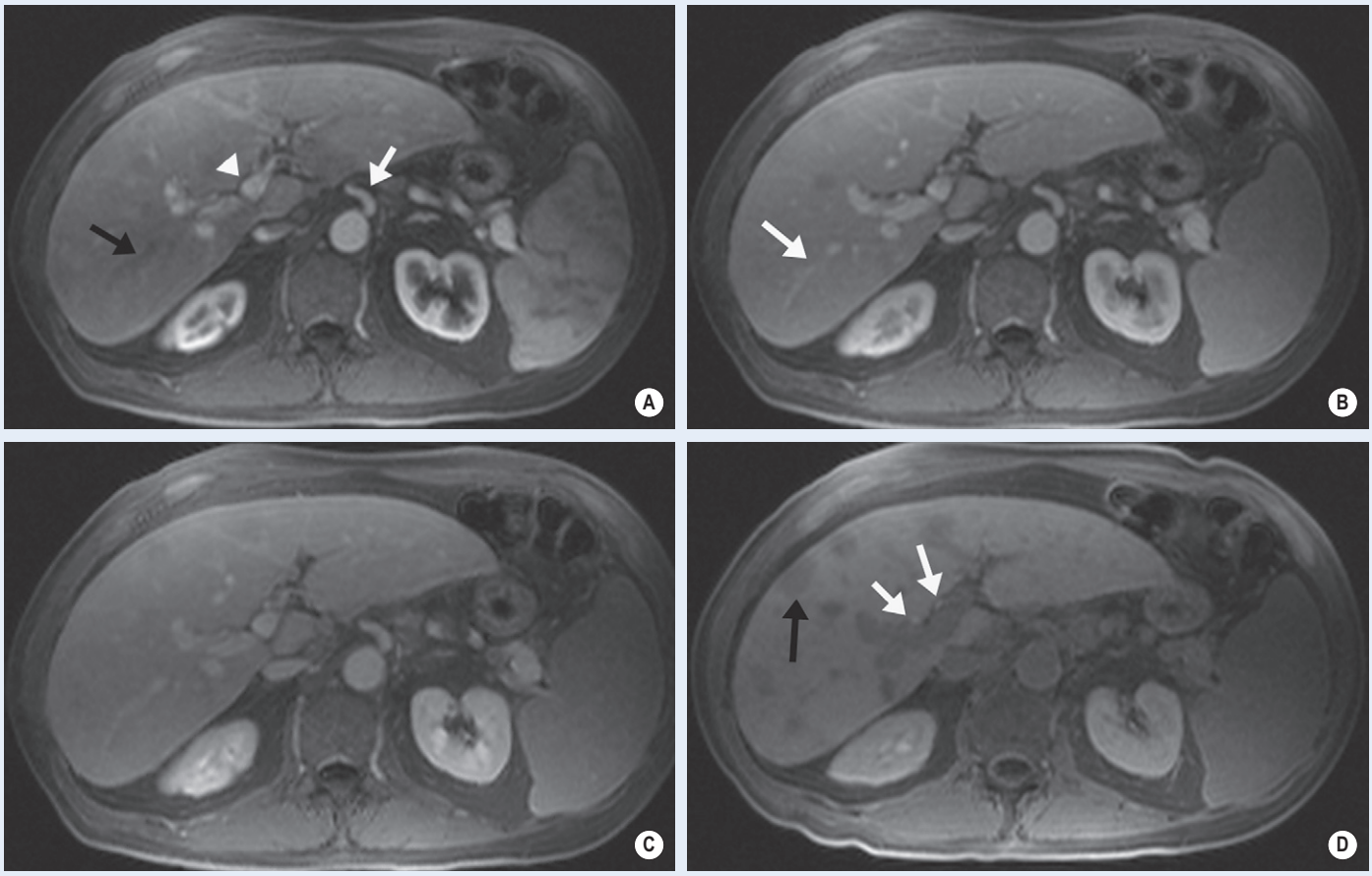
MRI CONTRAST AGENTS

- MR imaging is an order of magnitude more sensitive to the effect of gadolinium than CT is to the effect of iodine – therefore a far lower dose of gadolinium is required for MR imaging
- Discontinue breast feeding for 24 hours following gadolinium ► only use gadolinium if absolutely necessary in pregnancy
- Extracellular MRI contrast agents do not cross the intact specialized vascular blood–brain barrier.
  - These agents accumulate in tissues with abnormal vascularity (malignant and inflammatory lesions) and in regions where the blood–brain barrier is disrupted
- Dynamic MR imaging of the liver is performed after bolus IV contrast injection ► fat-saturated three-dimensional (3D) volume interpolated MR imaging (e.g. VIBE, THRIVE, FAME) allows high spatial resolution imaging of the entire liver to be acquired in a 20-s breath hold
- Imaging is repeated in the arterial, portovenous and parenchymal phases of liver enhancement:
  - *Hepatic arterial phase:* typically 20–30 s after IV injection
  - *The portovenous phase:* 60–90 s after IV contrast medium administration
  - *Interstitial phase:* approximately 90 s to 5 min after administration of IV contrast agent
  - *Delayed hepatic enhancement phase:* this occurs at 15–30 min for Primovist and approximately 1–3 h for MultiHance
- **Hepatocyte selective gadolinium chelates:** paramagnetic compounds that are taken up by functioning hepatocytes and excreted in bile ► T1WI: increased SI
  - A major indication for their use is to characterize lesions as hepatocellular or non-hepatocellular:
    - *Hepatocyte-containing masses* (e.g. FNH): usually enhance
  - *Adenoma:* they appear hypointense as they do not contain normal biliary radicals
  - *FNH:* hyper- or isointense
  - *Metastases (non-hepatocellular) and hepatocellular carcinoma (poorly functioning hepatocytes):* frequently hypointense
- **Non-gadolinium contrast media:** unlike the gadolinium-based contrast media, dynamic imaging is not performed
  - Teslascan is infused intravenously and its selective uptake by hepatocytes results in intense T1 liver signal enhancement at approximately 30 min, persisting for several hours
- **Reticuloendothelial system iron-based contrast media:** iron accumulates within Kupffer cells in the normal liver, resulting in reduction in signal intensity of the liver during T2*-weighted gradient-echo imaging ► lesions containing Kupffer cells demonstrate signal reduction, whereas lesions that are Kupffer cell depleted remain high signal
  - They are used most routinely to aid in HCC detection in high-risk patients ► HCC detection in cirrhosis with gadolinium may be difficult due to fibrosis, regenerating nodules and altered liver perfusion
  - NB: well-differentiated HCC may accumulate SPIO particles

Nephrogenic systemic fibrosis (NSF)

- The disease is characterized by scleroderma-like skin changes mainly affecting the limbs and trunk – this can progress to flexion contracture of joints ► the fibrotic changes may also affect other organs such as muscles, heart, liver and lungs
  - NSF is associated with gadolinium use and thus contraindicated with severe renal impairment (GFR <30ml/min)
  - Use as low a dose as possible in moderate impairment (GFR 31-48ml/min)
  - Immediate haemodialysis is not protective
- The stability of the binding of the gadolinium ion (Gd^{+++}) within the chelate could be an important factor in the pathogenesis ► the stability of the Gd chelates is influenced by the configuration of the molecule (whether linear or macrocyclic as well as its ionicity)
  - Macrocyclic chelates offer better protection and binding to Gd^{+++} (cf. linear molecules) ► therefore the least stable molecules are the non-ionic linear chelates
    - *High risk*: Omniscan (linear – non-ionic) ► OptiMARK (linear – non-ionic) ► Magnevist (linear – ionic)
    - *Medium risk*: Primovist (linear – ionic)
    - *Low risk*: ProHance (cyclic – non-ionic)

Classes	Non-specific extracellular gadolinium chelates	Hepatocyte selective gadolinium chelates	Non-gadolinium-based contrast media		
Examples	Magnevist ProHance Gadovist	MultiHance Primovist	Teslascan (manganese based)	Endorem (iron oxide based)	Resovist (iron oxide based)
Constituents	Low-molecular-weight gadolinium chelates	Low-molecular-weight gadolinium chelates: Gd-BOPTA, Gd-EOB-DTPA	Mangafodipir trisodium (MnDPDP)	Superparamagnetic iron oxide particles	Superparamagnetic iron oxide particles
Action	Distributes freely in the *extracellular* space	Initially distributes freely in *extracellular* space but undergoes *hepatic* excretion	Selective uptake by *hepatocytes* and excreted into bile ducts	Selective uptake by *Kupffer* cells	Selective uptake by *Kupffer* cells



MR study of the liver with a hepatocyte specific gadolinium chelate. (A) Arterial phase. There is contrast within the hepatic artery (white arrow). Contrast within the portal vein (arrowhead) indicates that sufficient time has passed for arterial enhancement of any lesion, but the lack of contrast within the hepatic veins (black arrow) ensures that there has not been any significant washout of contrast from the liver parenchyma. (B) Contrast is now seen within the hepatic veins (white arrow), indicating that a true parenchymal phase of enhancement has been reached. (C) Interstitial phase at about 3 minutes. (D) Delayed imaging with a hepatocyte specific agent, as indicated by contrast excretion via the biliary tree (white arrows). The multiple liver metastases that are hepatocyte poor and do not take up contrast are now much more visible on delayed imaging.

3.7 BILIARY

MAGNETIC RESONANCE CHOLANGIOPANCREATOGRAPHY (MRCP)

Technique

- Heavily T2-weighted coronal oblique fast spin-echo sequence to obtain source data (aligned along the plane of the common bile duct (CBD))
  - Stationary water appears as areas of high SI and adjacent soft tissue is low SI (therefore it is not reliant on contrast excretion and can be used in jaundiced patients)
  - Fasting reduces any unwanted signal from the adjacent intestine
  - Breath-hold or non-breath-hold (respiratory triggered) imaging
- Source data allows MIP reformats to be generated (highlighting fluid-filled structures) – usually a number of coronal MIP reformats over 180°
- *Secretin:* this stimulates exocrine pancreatic secretion, distending the pancreatic duct and improving its visualization (acts immediately, returning to baseline at 10 min)
- *Functional MR cholangiography:* using delayed imaging at 30–60 mins with the hepatobiliary excreted contrast agents Gd-EOB-DTPA (Primovist) or Gd-BOPTA (MultiHance)
  - *Uses:* liver donor transplant work-up ► the assessment of bile leaks and biliary communication with cysts ► the demonstration of segmental obstruction

Normal anatomy

- *Normal morphology:* only central intrahepatic ducts are normally seen ($\leq$ 3mm) ► extrahepatic ducts $\leq$ 7mm (CBD up to 10mm post cholecystectomy) ► pancreatic duct $\leq$ 3mm ► accessory pancreatic duct in 45%
- *Right posterior hepatic duct* (segments VI/VII): almost horizontal course
- *Right anterior hepatic duct* (segments V/VIII): more vertical course
- *Left hepatic duct* (segments II–IV): joins the right to form the common hepatic duct ► separate drainage of segment I
- *Cystic duct insertion into common hepatic duct:* right lateral (50%) ► anterior (30%) ► posterior (20%)
- *Common variants:* an aberrant right posterior duct draining into the common hepatic duct or cystic duct ► drainage of the right anterior or posterior duct into the left hepatic duct ► a triple confluence at the hilum

Imaging pitfalls

- *Technique:* volume averaging artefacts in MIP reformats can obscure filling defects – source images must always be reviewed ► MIP reformats can also over- and underestimate strictures
- *Normal variants:* a long cystic duct running parallel to the CBD, stimulating a distended CBD ► a contracted sphincter mimicking an impacted stone
- *Intraductal factors mimicking filling defects:* aerobilia (non-dependent) ► flow phenomena (central signal void) ► debris ► haemorrhage
- *Extraductal factors:* pulsatile vascular compression from adjacent vessels mimicking a stricture (but no proximal dilatation) ► susceptibility artefact from surgical clips

HEPATOBILIARY SCINTIGRAPHY

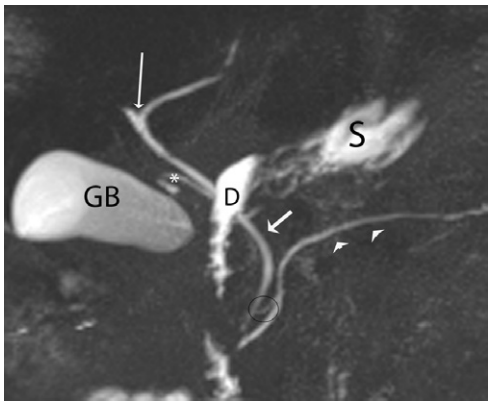
- Hepatobiliary iminodiacetic acid (HIDA) scintigraphy: this is a bilirubin analogue labelled with ^{99m}Tc
  - It is injected intravenously with serial images obtained over 2–4 h (it requires near-normal bilirubin levels)
- There is normally accumulation of isotope within liver, bile ducts, gallbladder, duodenum and small bowel by 1 h
  - *Delayed hepatic activity:* hepatocellular disease (with corresponding elevated bilirubin levels)
  - *Non-demonstration of the gallbladder:* acute cholecystitis ► a contracted gallbladder (e.g. following a recent meal)
  - Drugs that may aid visualization:
    - *Cholecystokinin:* this contracts the gallbladder
    - *Morphine:* this causes spasm of the sphincter of Oddi, therefore distending the biliary tree

ENDOSCOPIC ULTRASOUND (EUS)

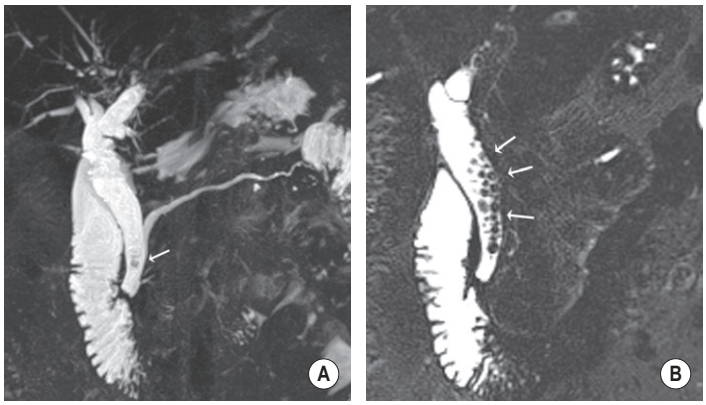
- This provides high-frequency grey-scale imaging (± colour Doppler) for the evaluation of the extrahepatic biliary tree, pancreas and duodenum ► it can also allow fine-needle aspiration cytology to be performed

ENDOSCOPIC RETROGRADE CHOLANGIOPANCREATOGRAPHY (ERCP)

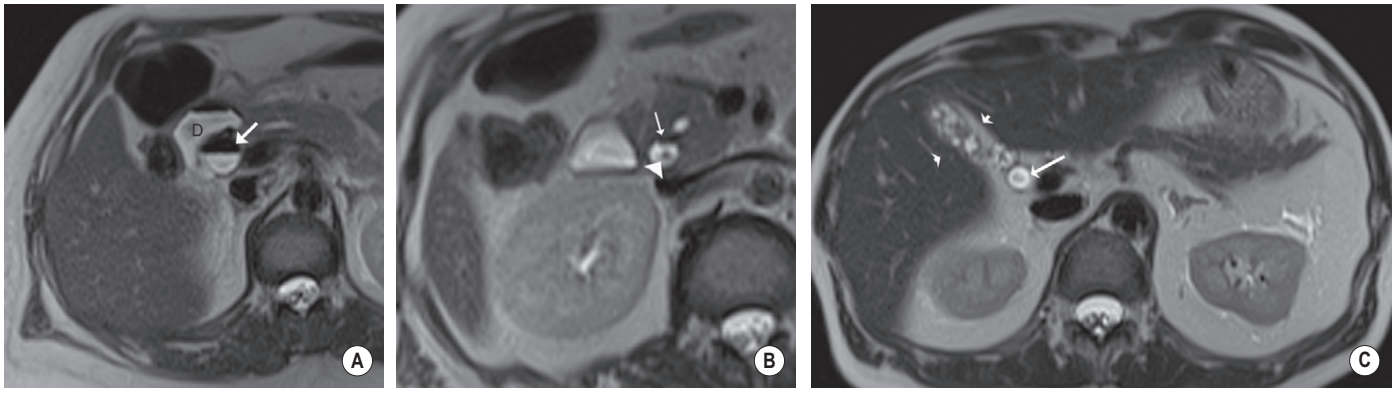
- This allows direct bile and pancreatic duct opacification, as well as visual assessment of the duodenum and ampulla of Vater
  - *It also allows for:* biopsy ► brushings ► sphincterotomy ► stone extraction ► biliary stenting ► biliary stricture dilatation
- The main complication is the precipitation of pancreatitis
- The main pitfall is the presence of underfilled ducts above a stricture



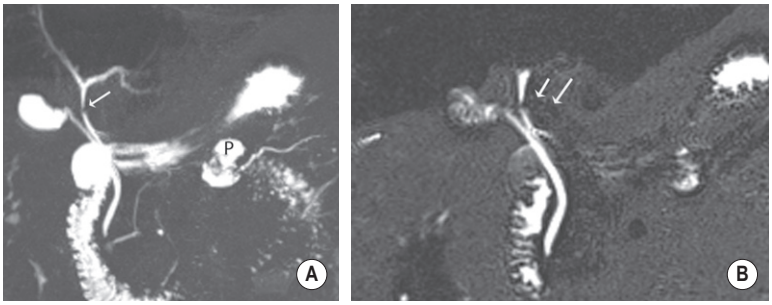
Normal anatomy on MRCP. The confluence of the right and left intrahepatic ducts to form the common hepatic duct is seen (long thin arrow). The cystic duct (*) typically joins the right side of the common hepatic duct to form the common bile duct (CBD) (short arrow). The main pancreatic duct (arrowheads) drains along with the CBD into the major duodenal papilla. An accessory pancreatic duct is present (black circle), draining into the minor duodenal papilla. Fluid containing structures such as the gallbladder (GB), duodenum (D) and stomach (S) are also clearly seen.



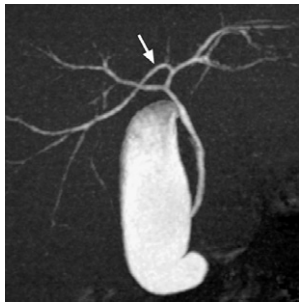
Example of a partial voluming artefact. (A) Coronal maximum intensity projection (MIP) reformat shows a possible filling defect (arrow) in the dilated distal CBD. (B) The thin section MRCP source image in fact demonstrates multiple filling defects (arrows) in the CBD, in keeping with stones.



Example of intraductal factors causing potential pitfalls in interpretation. (A) Axial T2-weighted MRI shows an air-fluid level in a dilated proximal CBD in keeping with aerobilia (arrow), adjacent to the duodenum (D), which also shows an air-fluid level. (B) More distally in the same patient, the cause of the obstruction is seen with a dependent filling defect (arrowhead) in the distal CBD in keeping with a stone. This should not be confused with the non-dependent aerobilia also shown at this level (arrow). (C) Axial T2-weighted MRI in a different patient shows a central filling defect in a dilated CBD which is due to flow artefact (arrow). The patient also has chronic cholecystitis with a contracted gallbladder (arrowheads).



(A) Coronal MIP reformat suggests a stricture or possible filling defect in the common hepatic duct (arrow) but with no upstream dilatation. Incidental note is also made of a small pseudocyst (P) associated with the main pancreatic duct. (B) Thin-section MRCP image more clearly shows that this is due to extrinsic compression from the right hepatic artery, which appears as a subtle curvilinear signal void outside the duct and extending across it (arrows).



Biliary duct anatomy. CT-IVC (maximum intensity reformat). Right posterior sectoral duct (arrow) passes to the left to drain into left hepatic duct.*

CHOLELITHIASIS

DEFINITION

- Stones present within the gallbladder – this affects 15% of the Western population (F>M) ► there is a small lifetime risk of developing a gallbladder carcinoma
- *Gallstone composition:* cholesterol (70%) ► pigment stones composed of calcium bilirubinate (up to 30%)

CLINICAL PRESENTATION

- Asymptomatic (80%) or presenting with biliary colic, acute or chronic cholecystitis, or obstructive jaundice

RADIOLOGICAL FEATURES

AXR Only 10–15% of calculi are visible (if they are calcified) ► larger stones tend to be laminated

US This has a sensitivity of > 95% for detecting gallstones ► gallstones appear as echogenic foci which cast acoustic shadows ► stone mobility is frequently demonstrated (unless it is impacted at the neck)

- NB: a gallbladder polyp will be fixed, with no acoustic shadow and may demonstrate vascularity

CT Only a minority of gallstones are visible ► these are hypodense, hyperdense or of mixed density

PEARLS

- **Reasons for non-visualization of the gallbladder:** a previous cholecystectomy ► a non-fasting state ► an abnormal gallbladder position ► emphysematous cholecystitis ► a gallbladder full of stones
- **Biliary sludge:** this is composed of calcium bilirubinate granules, cholesterol crystals and glycoproteins ► it is commonly see with fasting states, critically ill patients, pregnancy, and in those patients receiving total parenteral nutrition ► it resolves spontaneously in 50% of cases

US Fine, non-shadowing dependent echoes ► small gallstones can be difficult to detect if they lie within any sludge

- Sludge can be differentiated from a tumour by its mobility, lack of internal flow, and lack of an associated gallbladder wall abnormality
- Blood (haemobilia) and pus (empyema) can appear similar to sludge (the clinical setting aids the diagnosis)

CHOLEDOCHOLITHIASIS

DEFINITION

Choledocholithiasis

- Stones within the bile duct
  - *Primary* (10%): arising within the bile duct (pigment stones)
  - *Secondary* (90%): stones that have passed from the gallbladder into the bile duct

Hepatolithiasis

- Intrahepatic stone formation
  - This may occur with common duct stones but is more often associated with other pathologies: benign strictures ► primary sclerosing cholangitis ► recurrent pyogenic cholangitis ► Caroli's disease

CLINICAL PRESENTATION

- Right upper quadrant pain ► obstructive jaundice ► pancreatitis

RADIOLOGICAL FEATURES

US An intraductal echogenic focus needs to be demonstrated in both the longitudinal and transverse planes (± duct dilatation) ► a duct diameter < 4mm carries a high negative predictive value for choledocholithiasis (regardless of the gallbladder status)

- Conditions mimicking a stone:
  - *Intraductal gas:* this has a linear nature and will be mobile
  - *Haemobilia and sludge:* this produces more diffuse echoes than a stone
  - *Surgical clips:* these will lie outside the duct lumen
  - *Parasites:* e.g. hydatid membranes

EUS This is more sensitive than standard US (with a sensitivity and specificity > 90%)

NECT A ring density or soft tissue density within the bile duct and surrounded by bile (sensitivity 60–88% ► specificity > 95%)

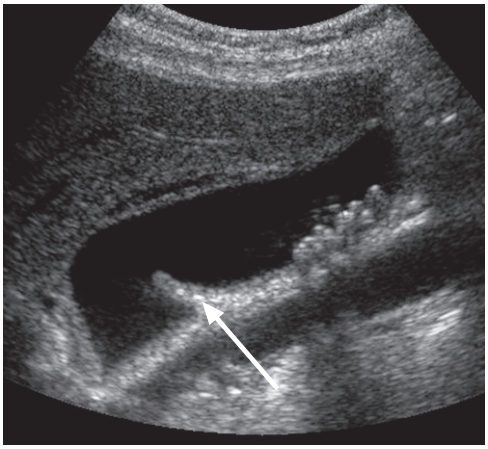
CT-IVC This has a high accuracy, with a reported sensitivity of up to 96% and a specificity of up to 98% ► it can diagnose stones that are <5mm in diameter ► its main weakness is its reliance on a near-normal serum bilirubin

MRCP An intraluminal signal void visible in 2 thin-section orthogonal planes ► this has a high sensitivity (up to 94%) and specificity (99%) ► its quality is independent of the serum bilirubin levels

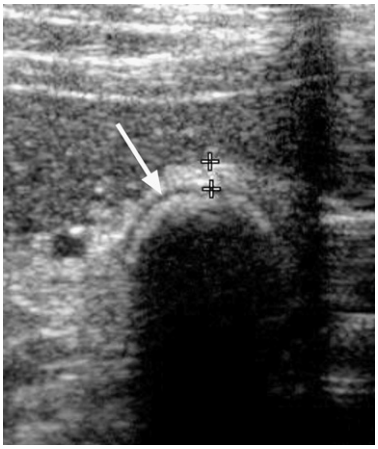
- False negative: stones <5mm
- False positive: gas (gas will rise, stones are dependent) ► haemobilia ► flow voids

PEARL

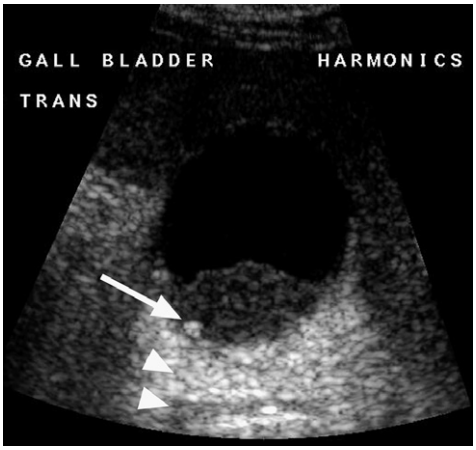
- 8–15% of patients who are under the age of 60 years and who have undergone a cholecystectomy have duct stones



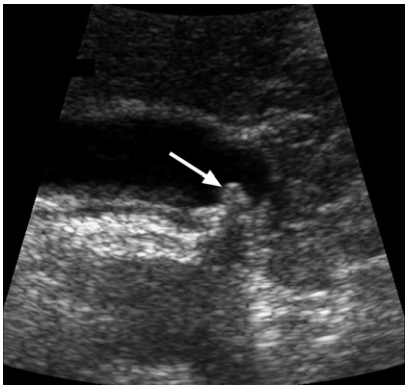
US shows multiple small shadowing stones. A normal fold (arrow) lies near the gallbladder neck.*



Gallbladder filled with stones producing the 'double-arc' sign ► hypoechoic line between two echogenic lines (arrow).*



Sludge within which a small stone (arrow) casts a subtle acoustic shadow (arrowheads).*



Choledocholithiasis. Small shadowing stone (arrow) in a dilated bile duct.*

Choledocholithiasis. A distal common bile duct stone (arrow) is slightly dense compared with the surrounding low-density bile.*

Choledocholithiasis. Single common duct stone (arrow) on thick-section, oblique, coronal MRCP. There has been a previous cholecystectomy.*

Choledocholithiasis. CT-IVC shows a small stone within the opacified distal common bile duct.*

ACUTE CALCULOUS CHOLECYSTITIS

Definition Gallbladder inflammation (which is secondary to gallstones in 90–95% of cases)

US This is the best initial imaging modality ► the signs include:

- A gallbladder wall thickness >3mm ► gallbladder distension (>5cm) ► pericholecystic fluid and gallbladder wall striations (± wall hyperaemia on Doppler) ► gallstones (common bile duct stones are suggested by abnormal liver function tests)
  - Fine echoes within the gallbladder may suggest the presence of sludge or pus (a gallbladder empyema)

CT Gallbladder wall thickening (>2mm) ► subserosal oedema and gallbladder distension ► high-density bile ► pericholecystic fluid and inflammatory stranding within the pericholecystic fat ► variable enhancement of the gallbladder wall

- Gallstones are only seen in a minority (as they are often isoattenuating to biliary fluid)

Hepatobiliary scintigraphy There is non-visualization of the gallbladder at 2–4 h after isotope administration (secondary to inflammatory cystic duct obstruction)

- **Complications:** Gangrenous cholecystitis ► emphysematous cholecystitis ► empyema formation
- **Differential of gallbladder wall thickening:** a non-fasted or a generalized oedematous state ► hepatitis ► pancreatitis ► gallbladder wall varices ► adenomyomatosis ► gallbladder carcinoma

GANGRENOUS CHOLECYSTITIS

Definition Ischaemic necrosis of the gallbladder wall is a complication of acute cholecystitis

Radiological features

US Irregularity or asymmetrical thickening of the gallbladder wall ► internal membranous echoes resulting from sloughed mucosa ► pericholecystic fluid

CT Gas within the wall or lumen ► discontinuous (±) irregular mucosal enhancement ► internal membranes (representing sloughed mucosa) ► a pericholecystic abscess

- **Gallbladder perforation:** this is seen in 5–10% and is suggested by pericholecystic fluid and localized gallbladder wall disruption

EMPHYSEMATOUS CHOLECYSTITIS

Definition The presence of intramural (± intraluminal) gas due to gas-forming organisms ► it accounts for 1% of cases of acute cholecystitis, and has a relatively high mortality rate

- 50% of patients are diabetic (M>F) ► gallstones are only seen in <50% of patients

US Focal or diffuse bright echogenic lines (representing intramural gas) ► a curvilinear brightly echogenic band with acoustic shadowing seen within a non-dependent portion of the gallbladder (representing intraluminal gas)

- Small foci of intramural gas may cause ring-down artefacts and mimic adenomyomatosis

CT Intramural (± intraluminal) gas

ACUTE ACALCULOUS CHOLECYSTITIS

Definition Gallbladder inflammation in the absence of gallstones ► this is usually found in critically ill patients

- *Other causes:* prolonged fasting ► parenteral nutrition ► AIDS ► diabetes ► chemotherapy

US Gallbladder distension ► gallbladder wall thickening ► echogenic contents (± sloughed membranes or mucosa) ► pericholecystic fluid

- Gallbladder aspiration may aid the diagnosis ► localized gallbladder tenderness is a good predictive sign but it is difficult to assess

CHRONIC CALCULOUS CHOLECYSTITIS

Definition Chronic inflammation and thickening of the gallbladder wall which is secondary to gallstones

US/CT A contracted gallstone-containing gallbladder ► intramural epithelial crypts (Rokitansky–Aschoff sinuses)

CHRONIC ACALCULOUS CHOLECYSTITIS

Definition Unexplained biliary-type pain with no clear clinical, pathological or radiological criteria for diagnosis

US This may show gallbladder wall thickening (but no gallstones)

Cholescintigraphy This can assess the gallbladder contractility (following an IV infusion of cholecystokinin) ► an ejection fraction < 35% indicates gallbladder dysfunction

XANTHOGRANULOMATOUS CHOLECYSTITIS

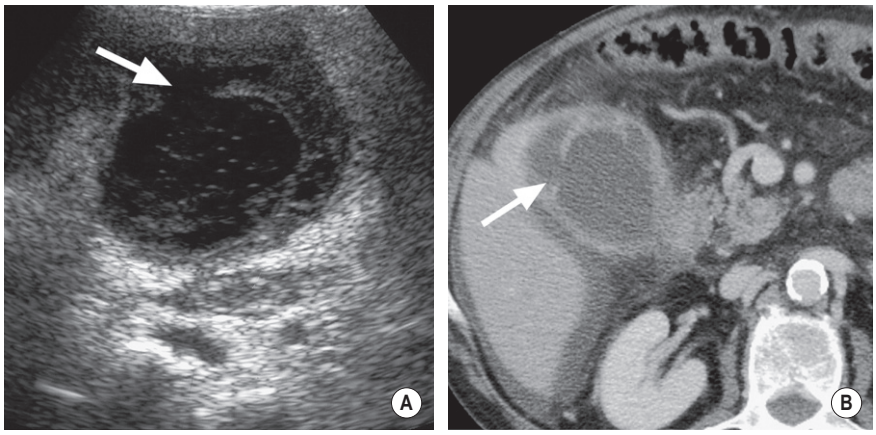
Definition A rare inflammatory disease of the gallbladder characterized by a focal, diffuse destructive inflammatory process with accumulation of lipid-laden macrophages ► it may simulate a malignancy radiologically and pathologically

Clinical presentation Cholecystitis or biliary obstruction

Radiological features

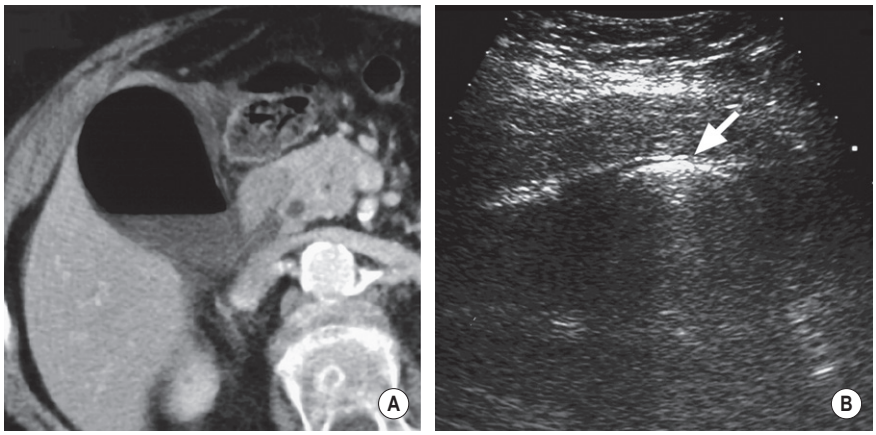
US/CT Gallbladder wall thickening (focal or diffuse) ► the majority have gallstones (± perforation, abscess, or fistula formation)

- An associated gallbladder carcinoma is seen in a minority of patients



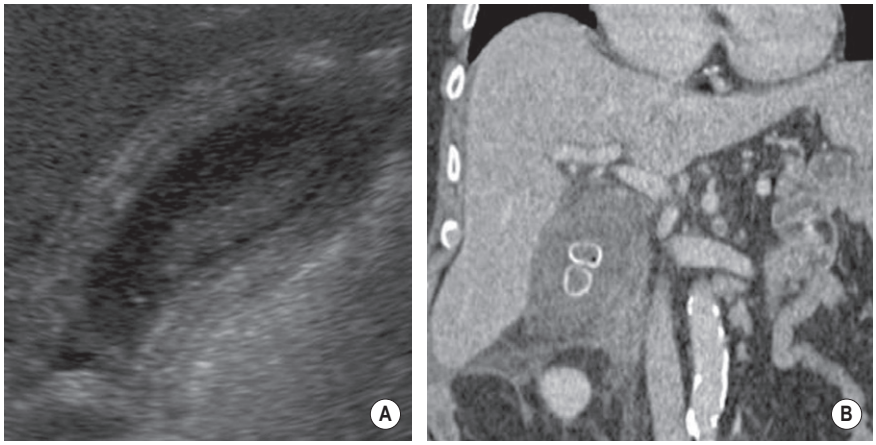
Acute cholecystitis with localized perforation on (A) US and (B) CT. The thickened gallbladder wall shows a local defect (arrow) and on CT there is small amount of intraperitoneal fluid and oedema of adjacent fat.*

Acute cholecystitis. The gallbladder contains small stones in the neck (Nos 1–4) and its wall shows oedematous thickening (5mm thickness).*

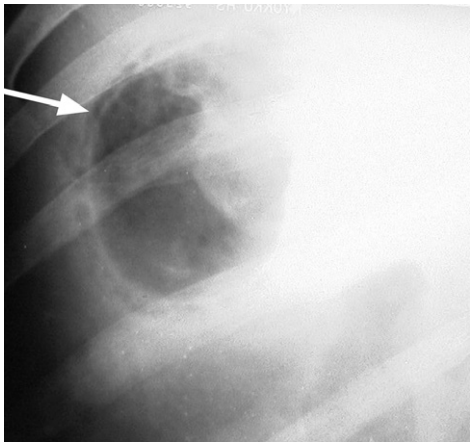


Emphysematous cholecystitis. (A) CT – intraluminal gas ► (B) US – intraluminal gas appears as a bright curvilinear echogenic band (arrow) with 'dirty' shadowing.*

Acute cholecystitis on CT. The gallbladder wall is thickened with oedema in the adjacent fat.*



Acute cholecystitis. (A). US demonstrating a thickened inflamed gallbladder wall. (B) Coronal CT demonstrating marked pericholecystic inflammatory stranding with laminated calcified gallstones in situ.



Emphysematous cholecystitis. Image showing intramural (arrow) as well as intraluminal gallbladder gas.*

ADENOMYOMATOUS HYPERPLASIA

DEFINITION

- This is otherwise known as adenomyomatosis or cholecystitis glandularis proliferans and is characterized by thickening of the gallbladder wall (resulting from epithelial and smooth muscle hyperplasia) ► it is associated with gallstones in 90% of cases
  - *Distribution:* fundal (the most common) ► segmental (usually within the mid-body) ► diffuse
  - The segmental form can lead to 'hourglass' deformity of the gallbladder
  - *Rokitansky–Aschoff sinuses:* cystic epithelial wall invaginations (which may contain small stones)

RADIOLOGICAL FEATURES

US Gallbladder wall thickening with secondary luminal narrowing ► the affected segment often contains wall bright echoes arising from the cystic spaces or from the small stones within them (and is often associated with 'comet-tail' ring-down artefacts)

CT Gallbladder wall thickening

MRI T2WI: intramural cystic spaces

CHOLESTEROLOSIS (STRAWBERRY GALLBLADDER)

DEFINITION

- This is due to cholesterol deposits within gallbladder wall macrophages ► it is associated with small polyps

RADIOLOGICAL FEATURES

US Echogenic foci within the gallbladder wall (with no acoustic shadowing)

GALLBLADDER FISTULAE

DEFINITION

- A rare condition due to either chronic stone disease (the majority) or neoplastic disease (the minority)
  - Cases due to chronic stone disease tend to fistulate with the duodenum ► cases due to neoplastic disease tend to fistulate with the colon
- *Cholecystoduodenal fistula:*
  - *'Gallstone ileus':* this is secondary to the antegrade passage of a gallstone and impaction within the terminal ileum
  - *Bouveret's syndrome:* this is secondary to the retrograde passage of a gallstone and obstruction within the stomach or duodenum

PORCELAIN GALLBLADDER

DEFINITION

- An asymptomatic and uncommon condition of mural wall calcification (focal or generalized) which is associated with chronic cholecystitis
  - Cholecystectomy is advocated as a carcinoma can occur in up to 30% of patients

RADIOLOGICAL FEATURES

US It may mimic an emphysematous cholecystitis

XR/CT Curvilinear calcification along the gallbladder wall

MILK OF CALCIUM BILE/LIMEY BILE

DEFINITION

Bile becomes very viscous, with a high concentration of calcium bilirubinate (due to stasis)

RADIOLOGICAL FEATURES

US Diffuse echoes similar to that seen with biliary sludge (but they are more echogenic with a tendency to layer out and produce an acoustic shadow)

CT/XR There may be layering of the high-density material

GALLBLADDER POLYPS

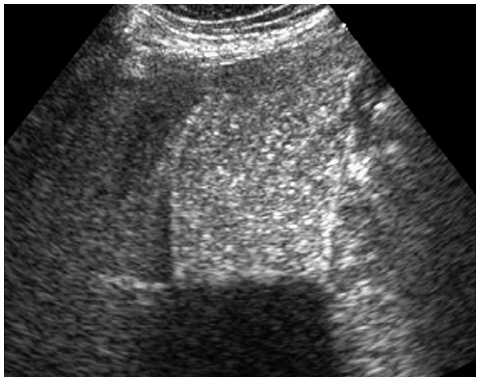
DEFINITION

- **Cholesterol polyps:** These account for the majority of polyps ► they are usually 2–10mm in size and are often multiple ► they are not usually associated with gallstones
- **Adenomatous polyps:** These are usually up to 2cm in size and are usually solitary ► they are often associated with gallstones
  - They are also associated with familial adenomatous polyposis and Peutz–Jeghers syndrome

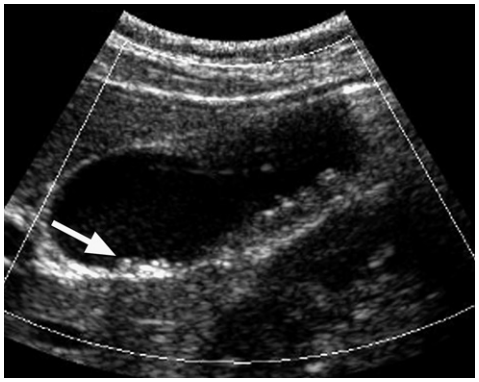
RADIOLOGICAL FEATURES

US Both types appear as small echogenic non-shadowing foci adherent to the gallbladder wall (often the non-dependent portion) ► they are usually non-mobile ► internal Doppler flow usually differentiates them from tumefactive sludge but this will not reliably distinguish between a benign and malignant polyp

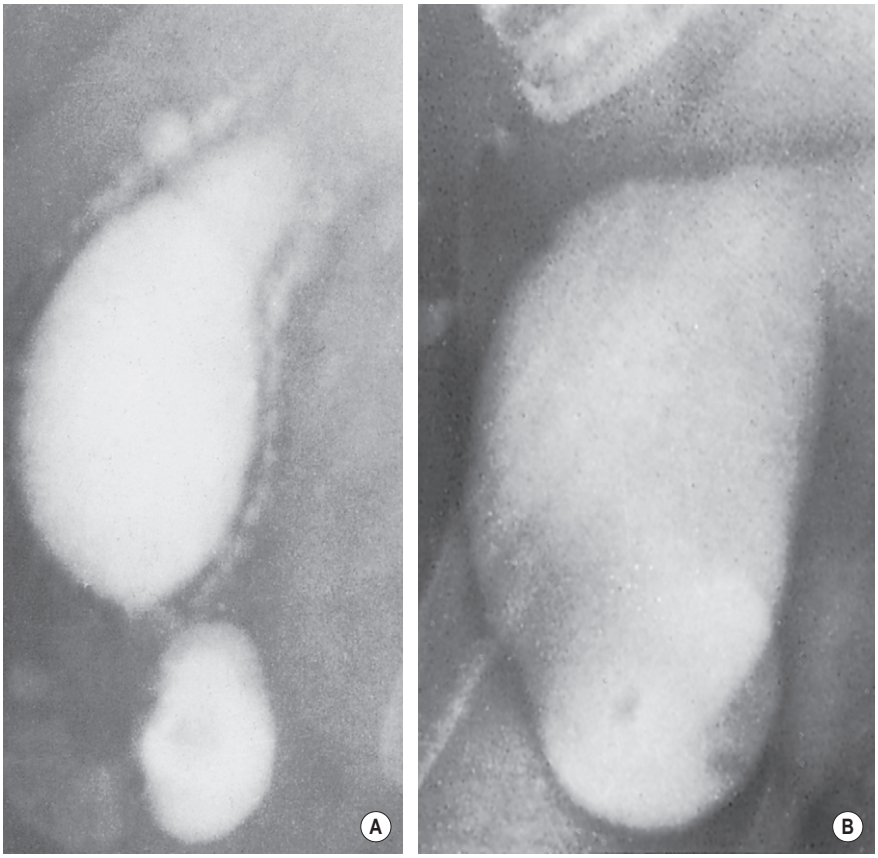
- A diameter of >10mm or local disruption of the adjacent gallbladder wall suggests malignancy



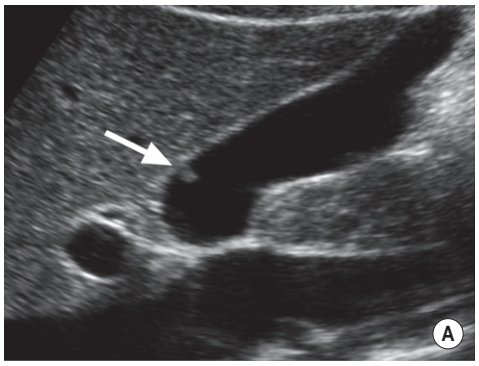
Milk of calcium bile producing fine echoes with a dependent layer that shadows.*

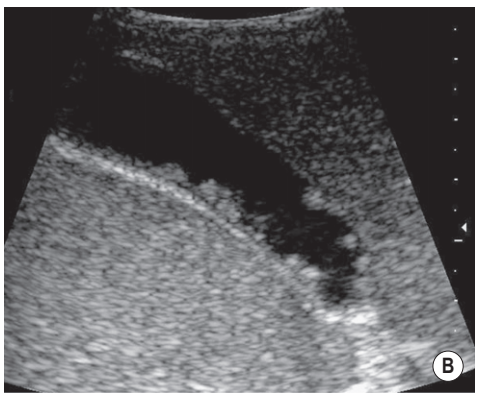


Adenomyomatous hyperplasia. Gallbladder wall thickening in the fundus is associated with small stones (arrow).*

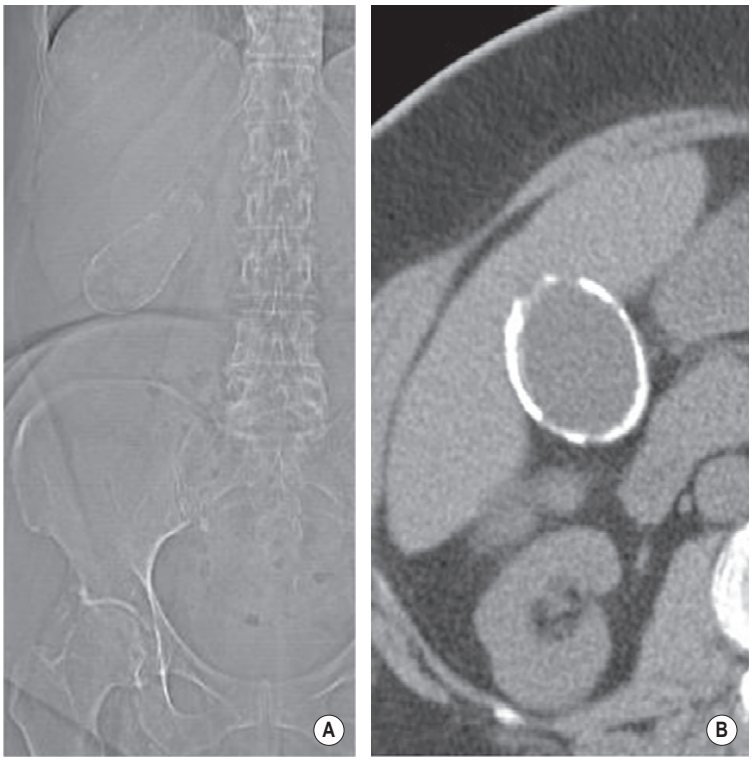


(A) Rokitansky–Aschoff sinuses shown on cholecystography. A stricture is also present. (B) Cholesterosis, showing fixed mural defects.†





Gallbladder polyps. (A) Solitary, non-dependent and non-shadowing polyp (arrow). (B) Multiple, non-shadowing cholesterol polyps.*



Porcelain gallbladder. NECT (A) and scout CT image (B).

POSTOPERATIVE STRICTURES

Definition A short (1–2mm) stricture usually seen following cholecystectomy ► they usually involve the common duct (as well as the hepatic and aberrant ducts) ► stones may develop proximal to a stricture

MRCP This can demonstrate the ducts above a complete stricture (unlike an ERCP)

- *Pitfall:* a common hepatic duct pseudostricture can be caused by the hepatic artery (or its right branch) crossing the duct

SCLEROSING CHOLANGITIS

Definition Inflammation of the intrahepatic (20%) and extrahepatic (80%) ducts ► it has an unknown aetiology

- *Primary disease (primary sclerosing cholangitis):* this is idiopathic
- *Secondary disease:* this is the most common form ► 70% of patients have a background of inflammatory bowel disease (usually UC)

Cholangiography Characteristic diverticula-like outpouchings alternating with strictures

- *'String of beads' appearance:* multiple segments of stricturing involving the intra- and extrahepatic ducts

US Bile duct wall thickening, which is most pronounced at the sites of stricturing ► outpouchings appear as local duct wall echogenic foci

CT/MRI Well-established disease is associated with areas of atrophy and hypertrophy within the liver

Bile duct stones (10%) These appear as high-density lesions on CT

Cholangiocarcinoma (10%) This should be suspected if there is progressive duct dilatation proximal to a stricture or if there is a nodule >1cm

MIRIZZI SYNDROME

Definition Chronic gallstone impaction within the gallbladder neck or cystic duct (or its remnant) leads to inflammation and fibrosis with associated common duct narrowing

- A fistula may develop between the gallbladder (or cystic duct) and the common duct – the stone may then partially or completely pass into the common duct

US Biliary dilatation down to a stone that is clearly not within the common duct

Cholangiography A smooth (2–3cm in length) stricture most commonly seen in the upper and middle common duct ► it often has a concavity toward the right

PANCREATITIS-RELATED STRICTURE

Definition Acute and chronic pancreatitis can produce biliary stricturing caused by fibrosis (± an inflammatory mass)

Cholangiography The strictures are smooth and tapering, extending over a few centimetres

HIV CHOLANGIOPATHY

Definition This typically occurs in patients with an established diagnosis of HIV and is due to opportunistic infection (most commonly *Cryptosporidium*)

US/cholangiopathy Bile duct wall thickening ► focal strictures (intrahepatic ± extrahepatic) ► biliary duct dilatation (which may be due to papillary stenosis) ► gallbladder wall thickening

ACUTE BACTERIAL CHOLANGITIS

Definition This is almost always caused by Gram-negative enteric organisms and is usually associated with at least partial bile duct obstruction (and is usually secondary to choledocholithiasis)

Clinical presentation Charcot's triad: fever + right upper quadrant pain + jaundice

Radiological features

US This can identify any duct stones as well as any bile duct wall thickening

Pearl Urgent imaging (US/CT/MRCP) is required to identify the cause and also for biliary tree drainage – this can be either endoscopic (ERCP and sphincterotomy) or transhepatic

RECURRENT PYOGENIC CHOLANGITIS/ ORIENTAL CHOLANGIOHEPATITIS

Definition Infection due to enteric bacteria or parasites (e.g. *Clonorchis sinensis*)

- This occurs mainly in South-East Asia or its emigrants ► it is characterized by recurrent episodes of cholangitis, biliary dilatation and strictures, together with bile duct stones

US Duct dilatation ► stones that may not shadow ► gas is often present within the ducts

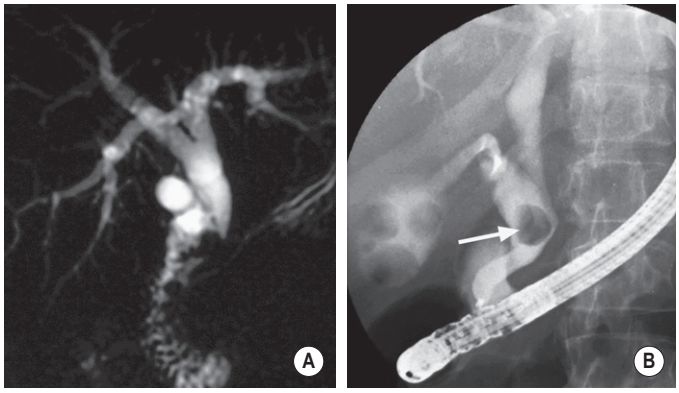
Cholangiography Duct dilatation and multiple duct stones (widespread or segmental) ► strictures

- *Clonorchis* is rarely identified as small filiamentous wavy or elliptical filling defects

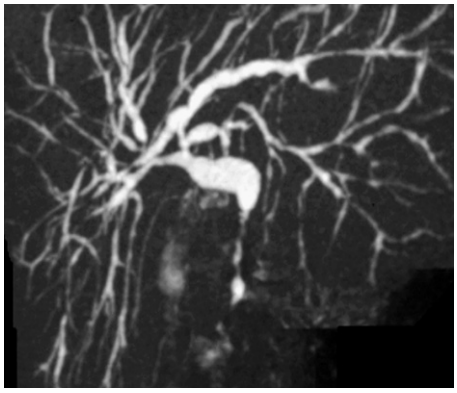
CT This identifies any associated hepatic abscesses and any lobar or segmental atrophy

- *Calcium bilirubinate stones*: these are seen within dilated ducts ► they are often intrahepatic and can be extensive

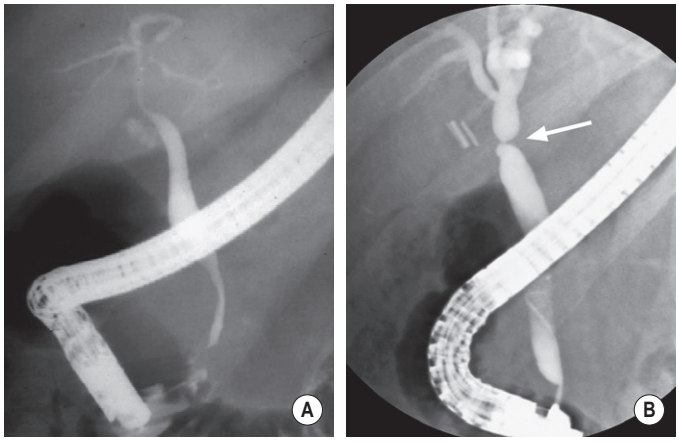
Complications Liver fibrosis ► portal hypertension ► cholangiocarcinoma



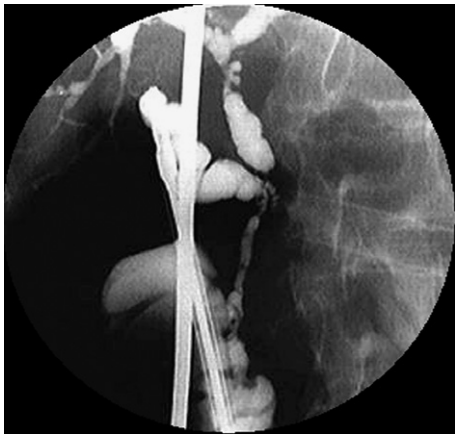
Mirizzi syndrome. MRCP (A) shows a stricture of the lower common duct caused by a stone (arrow) lying in an expanded cystic duct on ERCP (B). Multiple gallbladder stones are also seen.*



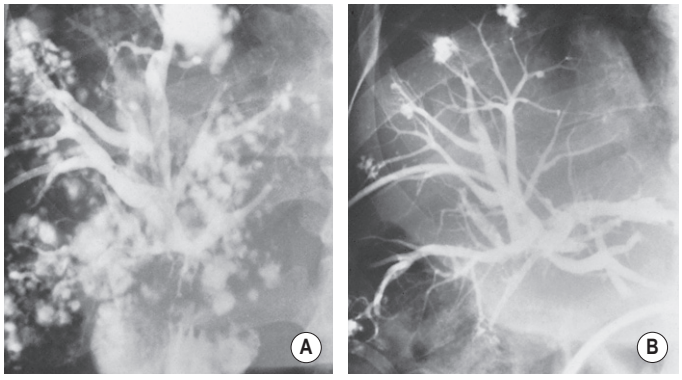
Primary sclerosing cholangitis. CT-IVC shows multiple intrahepatic and extrahepatic segments of stricturing + dilatation ('string of beads').*



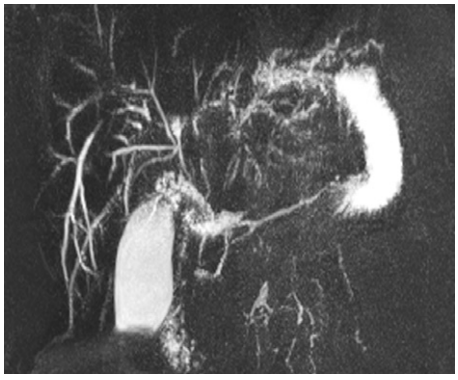
(A) Chronic pancreatitis. Typical smooth, elongated, incomplete stricture of the lower common bile duct. (B) ERCP shows a post-cholecystectomy stricture (arrow) which, characteristically, is very short.*



Primary sclerosing cholangitis. An intraoperative cholangiogram shows strictures and characteristic diverticula-like outpouchings affecting the common duct.*



Acute suppurative cholangitis. (A) Abscess cavities communicate with dilated ducts. (B) After 5 days of external drainage most of the abscess cavities have healed and the ducts are less dilated.†



MRCP showing multiple strictures and bile duct dilatation involving the intrahepatic ducts (especially the left-sided ducts) and the common bile duct in a patient with advanced HIV cholangiopathy.

CHOLANGIOCARCINOMA

Definition

- Adenocarcinomas originating from bile duct epithelium (> 95%)
  - ***Intrahepatic and peripheral to the liver hilum*** (10%) ► peripheral to the secondary bifurcation of the left or right hepatic ducts ► < 10% are diffuse or multifocal ► presents with abdominal pain/weight loss (obstructive jaundice is rare)
    - *Treatment:* hepatectomy
  - ***Hilar*** – Klatskin tumour (25%) ► arising from one of the hepatic ducts or the bifurcation of the common hepatic duct ► presents with obstructive jaundice
    - *Treatment:* bile duct resection + hepatectomy
  - ***Extrahepatic*** (65%): presents with obstructive jaundice
    - *Treatment:* pancreatoduodenectomy
- Pathologically 3 types:
  - ***Periductal infiltrating and stricture forming:*** the most common ► concentric mural thickening with stricture formation (± fibrotic encasement of adjacent vascular structures)
  - ***Mass forming (exophytic):*** frequent central necrosis/fibrosis (± satellite nodules)
  - ***Intraductal papillary growing:*** often small but cause obstruction, can secrete mucin, which tends to produce duct expansion (± calcification)
- **Risk factors:** primary sclerosing cholangitis (> ulcerative colitis) ► Caroli's disease ► choledochal cyst ► previous *Clonorchis* exposure (Asia) ► exposure to benzene or toluene

Radiological features

- **Intrahepatic**
  - CT/MRI: initial irregular peripheral patchy enhancement (central fibrosis) ► delayed progressive central in-filling ► capsular retraction due to fibrosis ► T1WI: hypointense ► T2WI: hyperintense
- **Hilar**
  - CT/MRI: biliary dilatation with left and right duct disassociation ► hilar bile duct wall thickening (relatively hypervascular or with delayed enhancement) ► any mass is usually small ► T1WI: hypointense ► T2WI: hyperintense
- **Extraheptic (distal CHD or CBD)**
  - CT/MRI: short stricture or polypoid mass ► thickened enhancing wall

Pearls

- **Hilar unresectability due to involvement of:** the secondary confluence (bilateral) ► main portal vein ► both portal vein branches ► hepatic artery + portal vein ► vascular involvement on one side of the liver and extensive biliary disease on the other
- **Metastatic spread:** this is commonly to the hepatoduodenal and portocaval nodes ► haematogenous remote spread is uncommon
- Intrahepatic tumours are staged as for a HCC
- MRCP is better than US and CT at evaluating the proximal extent of any stricturing (which critically affects the treatment options)

GALLBLADDER CARCINOMA

Definition An uncommon tumour (adenocarcinoma in 90%)

Clinical presentation There is usually a late presentation and consequently a very poor prognosis (unless it is detected incidentally at cholecystectomy) ► it presents during the 6th and 7th decades with RUQ pain (± biliary obstruction)

- *Risk factors:* cholelithiasis ► a porcelain gallbladder ► primary sclerosing cholangitis ► a choledochal cyst ► chronic infection
  - Any chronic inflammation will predispose to mucosal metaplasia

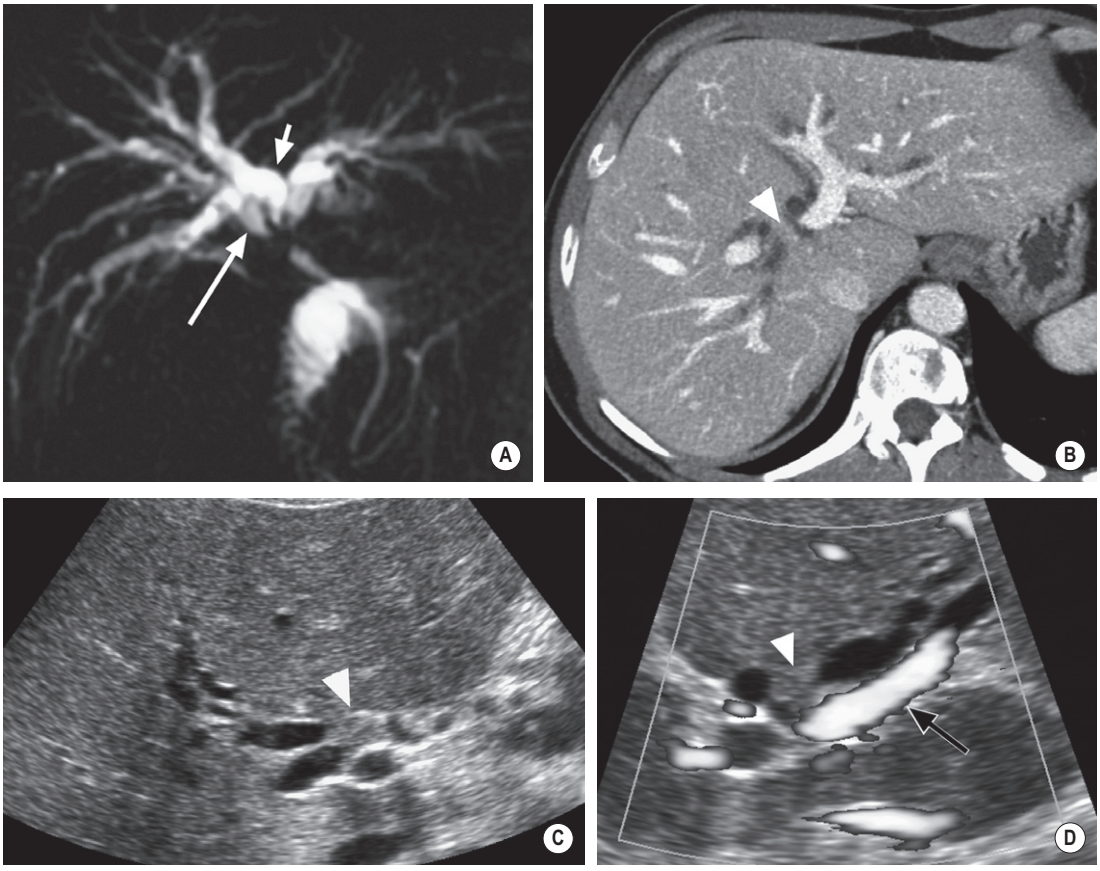
US/CT

- **Early (minority):** a polypoid intraluminal mass
- **Late (majority):** focal or diffuse irregular gallbladder wall thickening ► a large vascular mass within the gallbladder fossa with little or no gallbladder lumen identifiable (± central necrosis within larger lesions) ► biliary obstruction ► gallbladder stones are usually buried within the mass
  - There is early spread to the periportal lymph nodes (a nodal mass can extend down to the head of the pancreas) ► it can also spread to the adjacent liver (commonly involving segments IV and V)

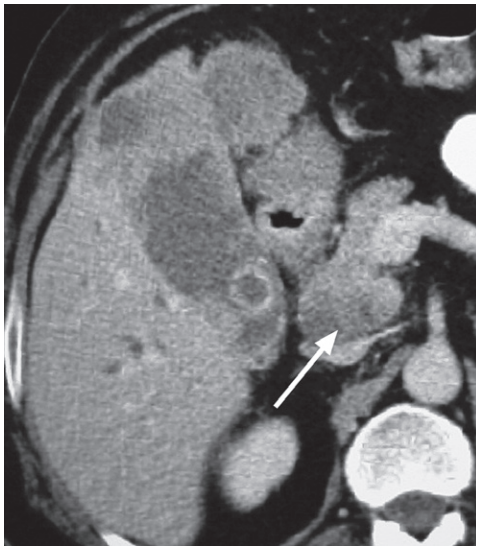
MRI T1WI: low SI ► T2WI: high SI ► T1WI + Gad: poor enhancement

Differential Mirizzi syndrome ► gallbladder metastases ► adenomyomatosis ► xanthogranulomatous cholecystitis

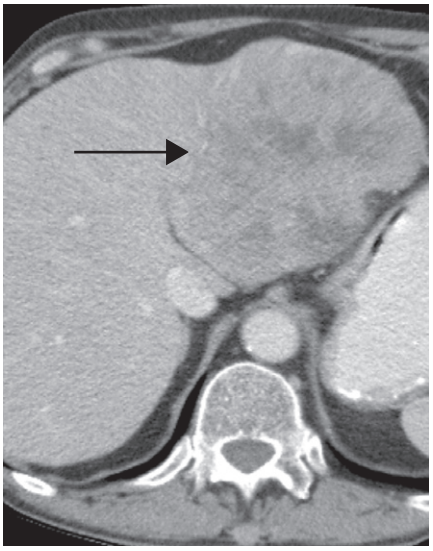
Bismuth-Corlette classification of extrahepatic biliary strictures	
Type I	Below the confluence of left and right hepatic ducts
Type II	Extension to the confluence of left and right hepatic ducts
Type IIIa	Type II + right hepatic duct involvement
Type IIIb	Type II + left hepatic duct involvement
Type IV	Involving both hepatic ducts or multifocal involvement



(A) Thick section oblique coronal MRCP. Small hilar cholangiocarcinoma (arrowhead) producing obstruction of the right posterior sectoral duct (short arrow), right anterior sectoral duct (long arrow) and left hepatic duct. (B) Axial portal phase CT. The small tumour is indicated by the arrowhead. (C) Longitudinal US. Again the tumour is indicated by the arrowhead. (D) Transverse colour Doppler US (black arrow: normal left portal vein).



Gallbladder carcinoma. Advanced carcinoma extending outside the fundus, with a nodal metastasis posterior to the pancreatic head (arrow). An associated stone can be seen in the gallbladder neck.*



Portal venous phase CT demonstrating a large heterogeneous cholangiocarcinoma occupying the left lobe of the liver (arrow).

BILIARY ATRESIA

Definition

- A progressive obliterative inflammatory process, affecting the extrahepatic biliary tree and progressing centrally towards the intrahepatic interlobar ducts
- The aetiology is uncertain: perinatal and embryonic, environmental, infectious, immune and genetic aetiologies have been suggested
- *Associated abnormalities occur in 10% of patients:* a preduodenal portal vein ► a choledochal cyst ► an absent IVC ► polysplenia and asplenia ► trisomy 13 ► situs inversus

Clinical presentation

- Persistent jaundice in the neonatal period (F>M) ► pale stools ► hepatomegaly

Radiological features

US The liver becomes large and coarse, with increased periportal reflectivity ► the biliary tree is not usually distended or dilated (due to the obliterative inflammatory process)

- The gallbladder is usually absent or rudimentary (it is seen in only 20%)
- *'Triangular cord' sign:* a highly reflective focus at the liver hilum cranial to the portal vein (representing the obliterated fibrosed biliary tree) ► if it persists after a Kasai procedure it may be a poor prognostic indicator
- The presence of the triangular cord sign or an absent or rudimentary gallbladder are high predictors of biliary atresia

^{99m}Tc-DISIDA This distinguishes biliary atresia from severe cholestasis

- ^{99m}Tc-DISIDA is extracted by the hepatocytes and secreted into the bile canaliculi and then into the bowel ► sequential imaging is performed during the first 60 min with delayed images taken at 2, 4, 6, and 24 h
- *Biliary atresia:* extraction is often normal with hepatic activity seen by 5 min ► failure to show excretion at 24 h (despite good parenchymal extraction) is suggestive of biliary atresia
  - The differential includes severe hepatocellular dysfunction

Preoperative PTC/PTTC and MRI These may play a role in difficult cases

Cholangiogram This is traditionally performed at laparotomy with a needle placed in the gallbladder or its remnant

Pearls

- The presence of a normal-sized gallbladder, which distends with fasting and contracts with feeding, suggests a diagnosis other than biliary atresia
- Percutaneous liver biopsy is required for a definitive diagnosis – this will show bile duct proliferation, periportal fibrosis, bile plugs and cholestasis

Treatment

- *Porto-enterostomy (Kasai procedure):* a jejunal loop is brought up as a Roux-en-Y up to the excavated porta hepatis to allow bile to drain through minute bile remnants or canaliculi into the bowel ► it is the primary treatment in patients who present before 60 days
- *Liver transplantation*: there is a significant morbidity and it requires long-term immunosuppression ► it is ultimately required in 70–80% of patients

Late complications These can occur even after a successful Kasai procedure: cirrhosis ► portal hypertension ► varices ► splenomegaly ► ascites

Other causes of neonatal jaundice Physiological jaundice of prematurity ► breast milk jaundice ► ABO incompatibility and other causes of haemolytic jaundice ► sepsis of any cause ► metabolic causes (e.g. galactosaemia, α_1-antitypsin deficiency and cystic fibrosis)

- *Unconjugated hyperbilirubinaemia:* this is caused by prehepatic and hepatic forms of liver disease
- *Conjugated hyperbilirubinaemia* (which is almost always pathological): includes extrahepatic obstructive forms (e.g. biliary atresia, Alagille syndrome, biliary hypoplasia and choledocal cysts) and hepatic forms (e.g. TPN and cholestasis)

BILIARY HYPOPLASIA (ALAGILLE SYNDROME)

Definition

- A paucity in the number of intralobular bile ducts
  - *'Non-syndromic':* this presents as an isolated finding
  - *'Syndromic'*: this was previously known as arteriohepatic dysplasia or Alagille syndrome

Clinical presentation

- Jaundice (presenting later than biliary atresia)
  - *Syndromic form:* forehead bossing ► a pointed chin ► posterior embryotoxin of the eye ► butterfly vertebrae ► renal anomalies (hypoplastic or dysplastic kidneys and cystic disease) ► peripheral pulmonary branch stenoses

Radiological features

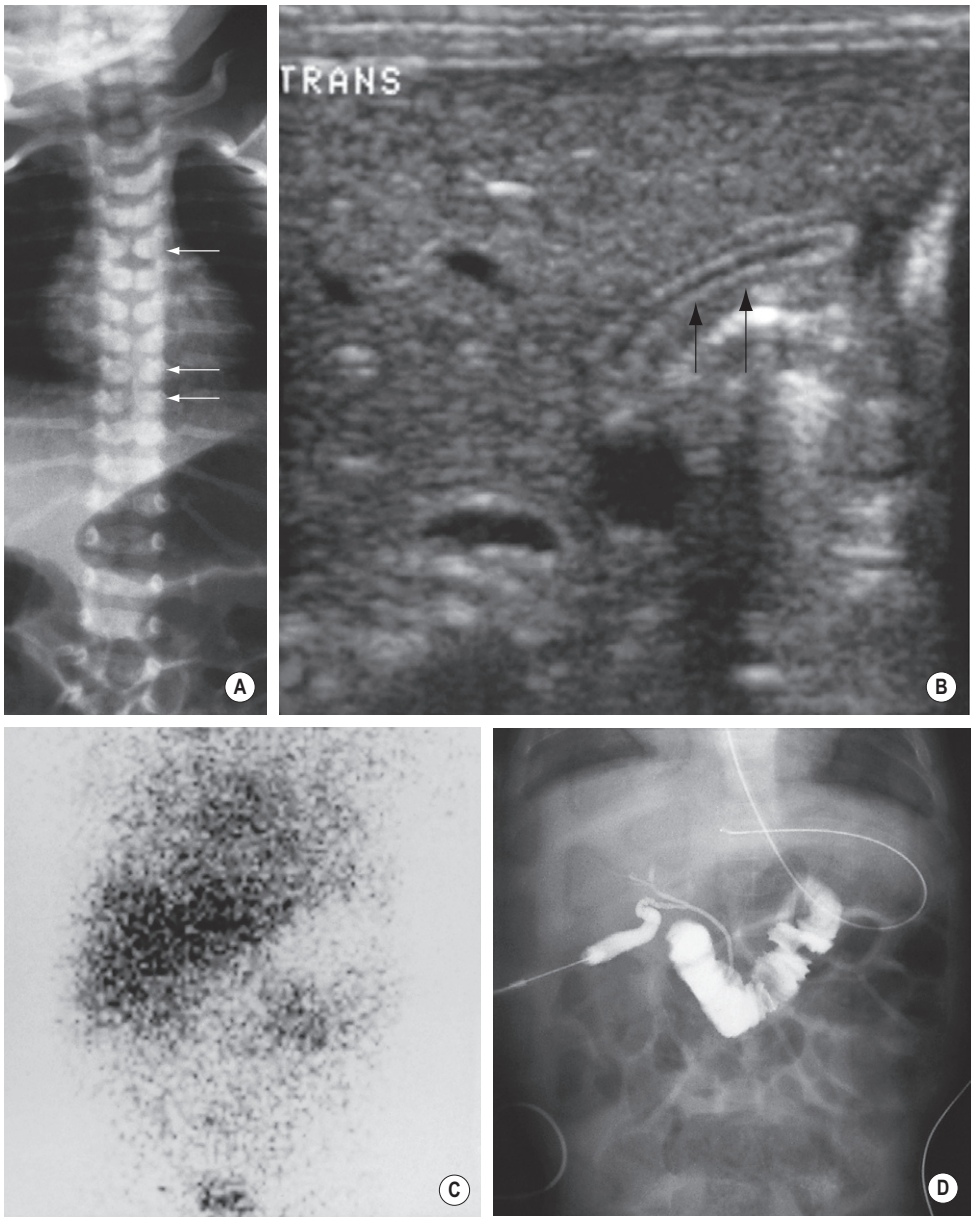
US A normal liver ► a normal or small gallbladder ► there is no triangular cord sign

^{99m}Tc-DISIDA No excretion into the bowel is seen in about 50% of biliary hypoplasias

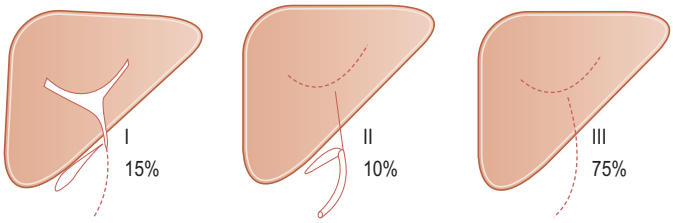
Cholangiography Patent thin spidery ducts

Pearls

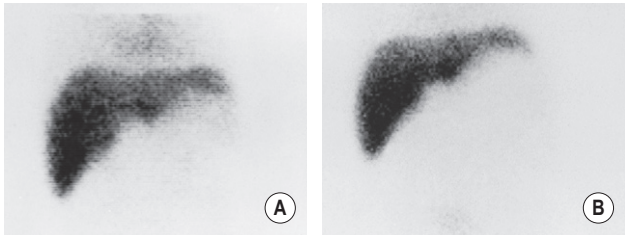
- The diagnosis is made on liver biopsy
- Management is conservative ► some complications may require liver transplantation
- *Late complications:* cirrhosis, portal hypertension and carcinoma



Alagille syndrome. Biliary hypoplasia. (A) Plain AP XR of the spine in a neonate with Alagille syndrome showing numerous hemivertebrae (arrows). (B) US of the liver in a 2-month-old boy with Alagille syndrome showing a small gallbladder (arrows). (C) Radionuclide scintigraphy in another child shows good extraction of the ^{99m}Tc-DISIDA by the liver, and excretion of some tracer into the bowel at 24 h. Both infants had biopsy-proven biliary hypoplasia. (D) Second image of a 2-month-old boy with prolonged neonatal jaundice. Preoperative cholangiogram shows a diminutive or hypoplastic but patent biliary tree consistent with biliary hypoplasia (non-syndromic). This was confirmed on biopsy.



Types of biliary atresia. Type I (extrahepatic) ► Type II (intrahepatic) ► Type III (combined)



Biliary atresia. Radionuclide study of a 2-month-old baby boy. ^{99m}Tc-DISIDA scintigram. Following preparation with phenobarbital for 5 days, this radionuclide scintigram using ^{99m}Tc-DISIDA shows good extraction of the tracer by the liver at 2 min, and no excretion by the biliary tree into the bowel by 6 h (A) or 24 h (B). Biopsy confirmed biliary atresia. Ultrasound showed no gallbladder present.*

HAEMOBILIA

Definition Most cases of bleeding into the biliary tree result from a blunt or penetrating trauma, or an iatrogenic injury (e.g. following liver biopsy)
- *Other causes:* hepatic artery aneurysms ► tumours ► cholecystitis

US Haemobilia appears similar to sludge in either the gallbladder or bile ducts

CT Slightly hyperdense material within the gallbladder and bile ducts

Cholangiography A cast-like filling defect within the bile ducts

BILIARY LEAKS AND BILE DUCT INJURIES

Definition This usually occurs following either a cholecystectomy or trauma

US/CT/MRCP These can detect biliary *collections*
- MRCP cannot usually identify the source of a leak

HIDA scintigraphy/CT-IVC/ERCP These can detect biliary *leaks*
- HIDA scintigraphy is the most sensitive technique
- ERCP: allows the placement of a temporary stent

High-quality cholangiography This is the most important imaging investigation following a major bile duct injury

BILIARY CYSTIC DISEASE (CHOLEDOCHAL CYSTIC DISEASE)

Definition A rare condition that is associated with biliary tumours (a 20-fold increase in cholangiocarcinoma) ► the Todani classification:
- *Type I:* dilatation (saccular or fusiform) of the common bile duct ► this is the most common type
- *Type II:* a diverticulum of the extrahepatic biliary duct
- *Type III:* a choledochocoele
- *Type IV:* multiple dilatations of the intra- and extrahepatic biliary tree ► this is the 2nd commonest type
  - Type 4a: fusiform dilation of the entire extrahepatic bile duct with extension of dilation into the intrahepatic bile ducts
  - Type 4b: Multiple cystic dilations involving only the extrahepatic bile duct
- *Type V*: Caroli's disease
- Types I and IV are characterized by a long common channel shared with the pancreatic duct ► there is a high incidence of stone development within the dilated ducts

Clinical presentation Type 1 cysts commonly present in childhood with pain, jaundice and a right upper-quadrant mass
- The presentation is otherwise similar to that seen with gallstone disease

ERCP, CT-IVC, PTC or contrast-enhanced MRCP These can be used ► cholangiography is the best test

CAROLI'S DISEASE

Definition A sporadic condition causing the formation of intrahepatic biliary cysts (with intrahepatic calculi) ► there is a risk of pyogenic cholangitis, intrahepatic abscess and cholangiocarcinoma formation
- *Associations:* medullary sponge kidney (80%) ► infantile polycystic kidney disease

US/CT A beaded appearance of the intrahepatic bile ducts with multiple cystic structures converging towards the porta in a branching pattern
- *'Central dot' sign:* a portal radical surrounded by a dilated bile duct

LIVER ATROPHY

Definition Lobar or segmental atrophy is frequently associated with contralateral lobar hypertrophy ► it may be associated with lobar or segmental bile duct obstruction due to malignant or benign causes
- *Malignant obstruction:* cholangiocarcinoma
- *Benign obstruction:* postoperative strictures ► primary sclerosing cholangitis

LOBAR OR SEGMENTAL DUCT OBSTRUCTION

Causes Stones ► postoperative strictures (which are usually right sided) ► primary sclerosing cholangitis ► cholangiocarcinoma

Clinical presentation
- A cholangitis or non-specific symptoms ► the serum bilirubin is usually normal but there is an elevated gammaglutamyl transferase and alkaline phosphatase

MRCP The best technique for evaluation

PARASITIC INFECTIONS: *ASCARIS LUMBRICOIDES*

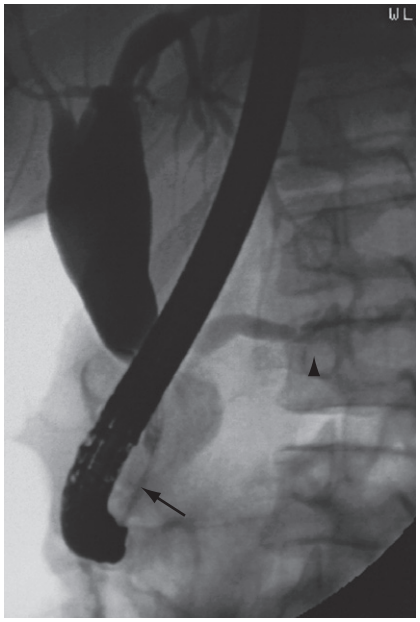
Definition A parasitic roundworm which enters the bile duct through a duodenal ampulla

Clinical presentation It can be asymptomatic or result in cholangitis, cholecystitis, or pancreatitis

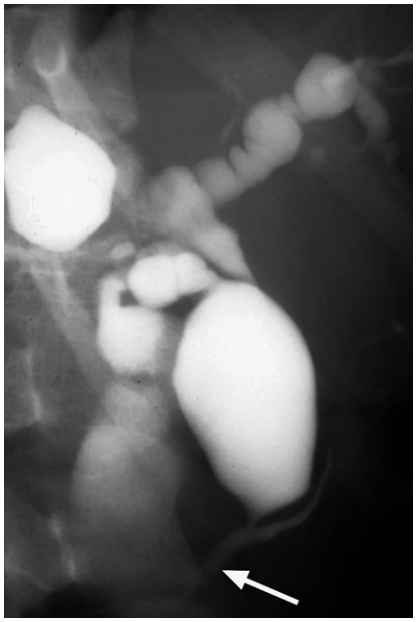
US/cholangiography A tube-like structure within the biliary tree

HYDATID DISEASE

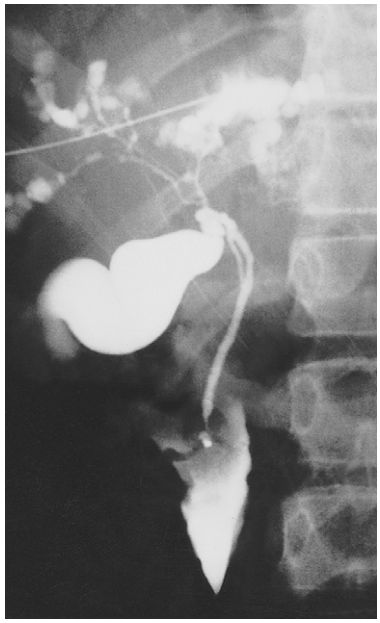
Pearl Hepatic hydatid cysts may rupture into the biliary tree (potentially obstructing it) ► ruptured membranes are seen as curvilinear structures on US/cholangiography



ERCP in choledochal cyst. Radiograph of an ERCP examination showing a moderate-sized fusiform choledochal cyst. Contrast medium fills the pancreatic duct (arrowhead) and the common channel is seen (arrow).*

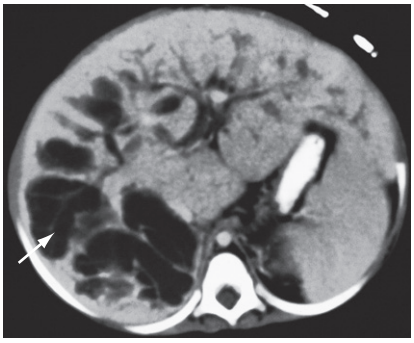


Type IV choledochal cyst on PTC with extrahepatic as well as intrahepatic cystic dilatation of bile ducts and a characteristic long common channel shared by the common bile duct and pancreatic duct (arrow).*

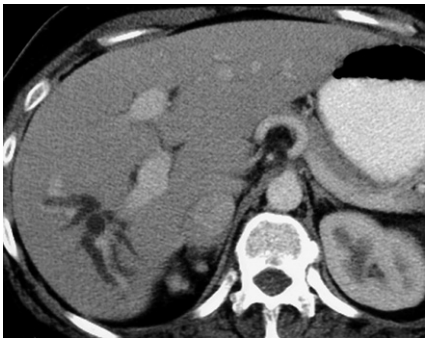


Caroli's disease with characteristic strictures and segmental intrahepatic dilated ducts.†

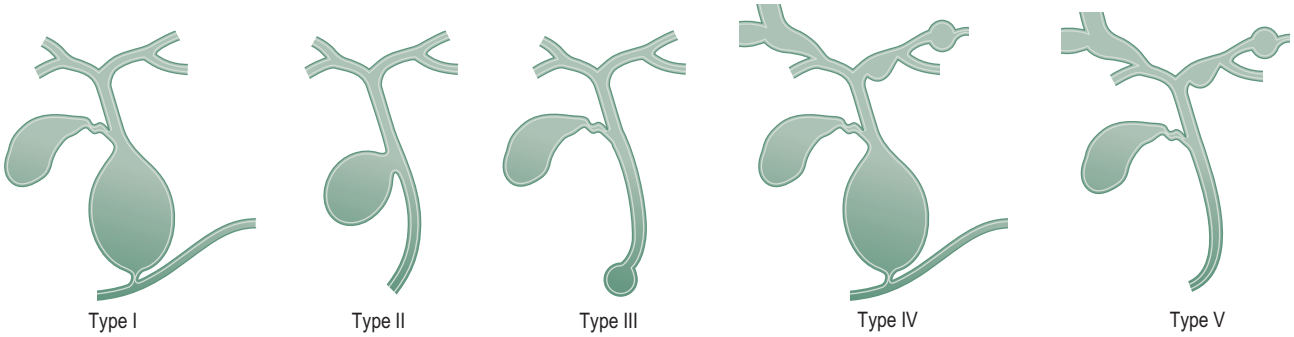
Hilar cholangiocarcinoma associated with marked right-lobe atrophy and left-lobe hypertrophy. MRCP shows a stricture with very dilated and crowded ducts in a small atrophic right lobe. Arrowhead = segment II ► long arrow = segment III (partially out of section).*



Caroli's disease. CT showing low attenuation areas surrounding the portal vein branches (arrow). The whole liver is involved, the right lobe more than the left.*



Right hepatic duct obstruction due to a postcholecystectomy stricture demonstrated on portal phase CT. The atrophic right lobe has rotated to lie in a characteristic posterior position. The left lobe is hypertrophic.*



Biliary cystic disease classification (after Todani).

PERCUTANEOUS CHOLECYSTOSTOMY

Indications

- *Acute calculous cholecystitis*: this is performed in patients who are a poor surgical risk because of their co-morbidities ► it may allow for a subsequent cholecystectomy once the patient has recovered or a percutaneous extraction of any gallbladder stones
- *Acalculous cholecystitis:* this is performed in critically ill patients

Technique

- A transperitoneal approach into the fundus of gallbladder, or a transhepatic approach into the body of the gallbladder (this avoids transgressing the peritoneum)
- If any stones cannot be cleared (and a cholecystectomy is not possible), the catheter can be removed after 2–3 weeks' drainage
  - A contrast study is first performed to show cystic duct patency and to exclude the presence of any bile duct stones requiring an endoscopic sphincterotomy
- With acalculous cholecystitis, the catheter is left in for at least 3 weeks

PALLIATION OF MALIGNANT BILIARY OBSTRUCTION

Endoscopic approach

- This can use either a plastic or metal stent ► this approach is preferred for a low biliary obstruction

Transhepatic approach

- This uses catheters or indwelling stents
- *Catheters:* these are inserted across malignant strictures in 80–90% of cases, establishing internal and external drainage
  - An *internal* stent is preferable to a *percutaneous* catheter as it avoids the ongoing discomfort and inconvenience associated with a percutaneous catheter
  - Percutaneous catheters can be used for brachytherapy access with ^{192}Ir, in combination with metal stent insertion ► this may improve stent patency and survival
- *Expanding metal stents:* these are preferable to *plastic stents* as they provide a much larger deployed lumen than that of the transhepatic track diameter ► they have longer patency rates, lower early complication rates and require fewer reinterventions
  - For strictures below the confluence of the right and left ducts the approach can be via the right lobe (through the right flank) or via the left lobe (through the epigastrium)
  - The right flank approach provides a less angled pathway at the biliary confluence and may result in less haemobilia

Hilar strictures: special considerations

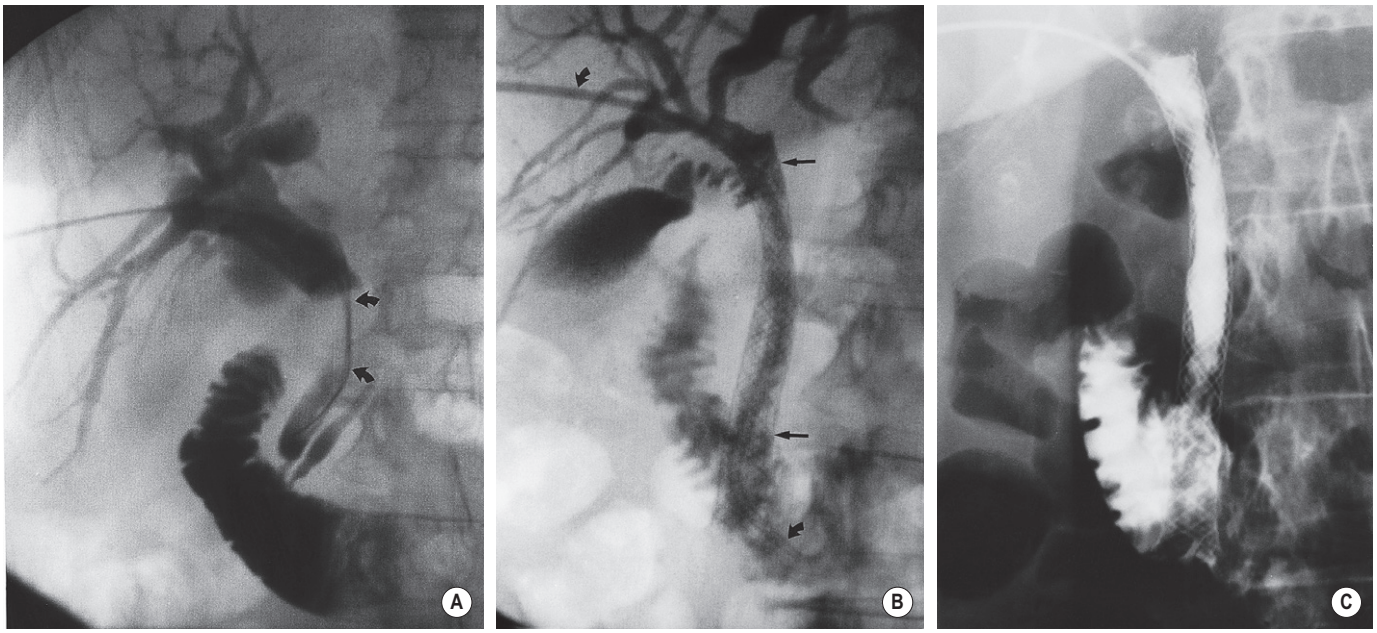
- A stent is placed to drain the largest possible volume of non-atrophic and tumour-free liver ► contralateral stenting is required if the opposite lobe contains infected bile in cases of suspected cholangitis (on FNA)

MANAGEMENT OF BENIGN BILIARY OBSTRUCTION

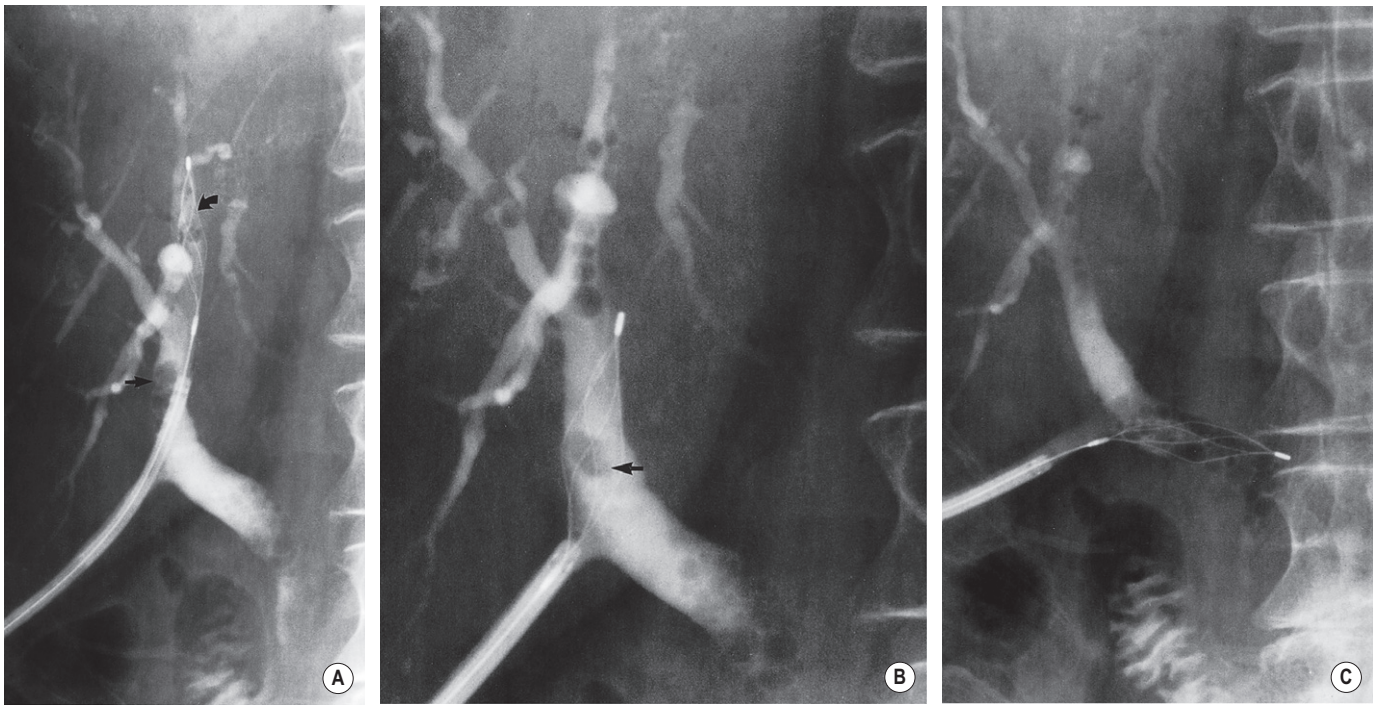
- Transhepatic procedures are indicated to:
  - Drain an obstructed infected system which is not amenable to endoscopic drainage
  - Dilate benign strictures when surgery or endoscopic treatment is not possible (a biliary drainage catheter needs to be placed across the stricture)
  - Treat intrahepatic or extrahepatic ductal stones that are not amenable to endoscopic or surgical management
- It may be combined with operative approaches for complex benign biliary problems

BIOPSY TECHNIQUES

- FNAC (22G needle) or biopsy (18G needle) of the bile ducts can be performed
- The total complication rate for FNAC of liver, biliary tract and pancreas is $< 0.2\%$ (e.g. bleeding, bile leak, pancreatitis and infection)
- In biliary obstruction, biopsy should be delayed until after there has been biliary decompression ► biopsy is not recommended if a patient is proceeding to an attempted resection
- Biopsy is usually image guided (e.g. US, CT or fluoroscopy with cholangiography) ► an ERCP allows for the direct biopsy of any tumours involving the ampulla or duodenum ► EUS may guide the biopsy of more distally obstructing tumours



Metal stent placement in a patient with pancreatic cancer who had a failed ERCP. (A) Biliary drainage was performed and the stricture (curved arrows) was traversed with a hockey stick catheter and hydrophilic guidewire. Note the normal duodenum without evidence of encasement. (B) 70 × 10mm Wallstent was placed across the stricture (straight arrows) and a safety catheter was left in situ for 3 days (curved arrows) because of some bleeding. (C) The patient returned for a tube injection 3 days later. The stent was in a good position with good flow of contrast into the duodenum. The safety catheter was removed.¶¶



Percutaneous extraction of a retained common bile duct stone through a T-tube track. (A) The T-tube has been removed and a steerable catheter placed through the percutaneous track into the bile duct to the level of the stone (small arrow). A basket (curved arrow) has been placed distal to the stone. (B) The basket is pulled back and manipulated so that the stone (small arrow) is engaged within the basket. (C) Basket, stone and catheter are then removed through the percutaneous track.¶¶

3.8 PANCREAS

EMBRYOLOGY

- The pancreas develops in two parts from the endoderm of the primitive duodenum
- **Dorsal part:** this is the first part to appear, initially appearing as a diverticulum from the dorsal wall of the duodenum ► it forms the neck, body and tail of the gland and part of the head
- **Ventral part:** this develops more caudally and initially appears as a diverticulum from the developing bile duct ► it forms the remaining part of the head and uncinate process
  - The duodenum undergoes partial rotation and the 2 parts approximate each other and fuse
    - Before this occurs the dorsal duct (the duct of Santorini) opens into the duodenum proximal to the major papilla (the ampulla of Vater)
    - The ventral duct (the duct of Wirsung) opens into the major papilla with the CBD
  - Usually fusion of the two ducts occurs at the junction of the head and body of the gland, with the ventral duct becoming the main excretory pancreatic duct (in >90% of cases)

CONGENITAL ANOMALIES

PANCREAS DIVISUM

Definition This follows failure of fusion of the dorsal and ventral ducts (affecting 5–10% of the population) ► it is the commonest congenital pancreatic anomaly ► it may result in functional stenosis and pancreatitis and there is an increased incidence of pancreatic malignancies

- The duct of Wirsung drains the head and uncinate process of the pancreas (via the major papilla)
- The duct of Santorini drains the body and tail (via the more cranially positioned minor papilla)

ERCP/MRCP These allow visualization of the ducts

ANNULAR PANCREAS

Definition Failure of normal rotation during development results in pancreatic tissue partially or completely encircling the duodenum ► this is the 2nd most common congenital anomaly

- It may cause proximal duodenal dilatation and symptomatic duodenal narrowing
- *Associations*: duodenal atresia and stenosis ► oesophageal atresia ► tracheo-oesophageal fistula ► Down's syndrome

Barium studies Narrowing of the duodenum at the level of the major papilla

CT Pancreatic tissue surrounding the duodenum

ERCP/MRCP These demonstrate a segment of pancreatic duct encircling the duodenum

PANCREATIC AGENESIS, HYPOPLASIA AND ECTOPIC PANCREAS

Definition Total pancreatic agenesis is rare – agenesis or hypoplasia of the dorsal part may occur

- Ectopic islands of pancreatic tissue may be found remote from the gland (e.g. within the gastric or duodenal wall)

Barium studies A smooth mural nodule, often with central umbilication (representing a rudimentary pancreatic duct)

MULTISYSTEM DISEASES WITH PANCREATIC INVOLVEMENT

CYSTIC FIBROSIS

Definition An autosomal recessive condition characterized by defects of serous and mucous secretion and involving multiple organs ► 85% of patients have severe exocrine pancreatic insufficiency and steatorrhoea

- Obstruction of the main pancreatic duct (and its side branches) by inspissated secretions results in acinar and ductal dilatation with subsequent atrophy of the acinar tissue

US/CT Marked fatty replacement of the normal pancreatic parenchyma ► dystrophic calcification ► pancreatic cysts

VON HIPPEL–LINDAU DISEASE

Definition An autosomal dominant condition characterized by renal cell carcinomas, phaeochromocytomas, retinal angiomatosis and cerebellar haemangioblastomas

Pearls The most common pancreatic lesions are simple pancreatic cysts ► serous cystic pancreatic neoplasms and pancreatic islet cell tumours may also occur

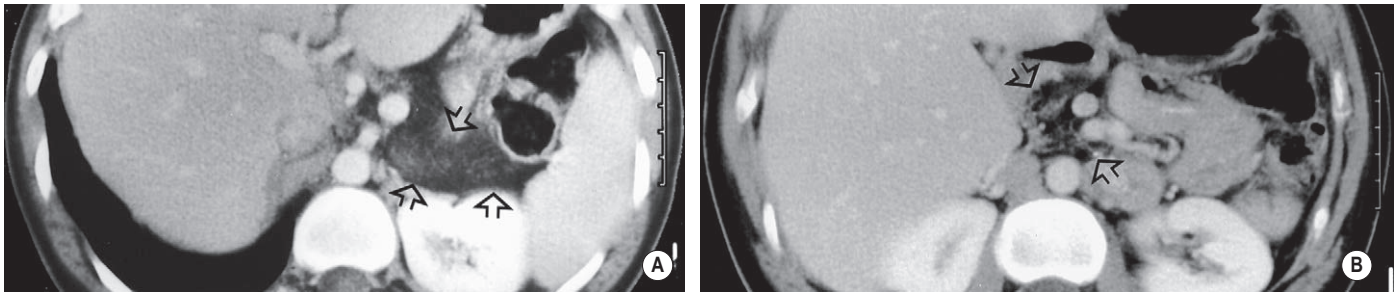
POLYCYSTIC KIDNEY DISEASE

Definition An autosomal dominant condition characterized by multiple renal cysts ► hepatic cysts may also occur and pancreatic cysts are seen in 10% of patients

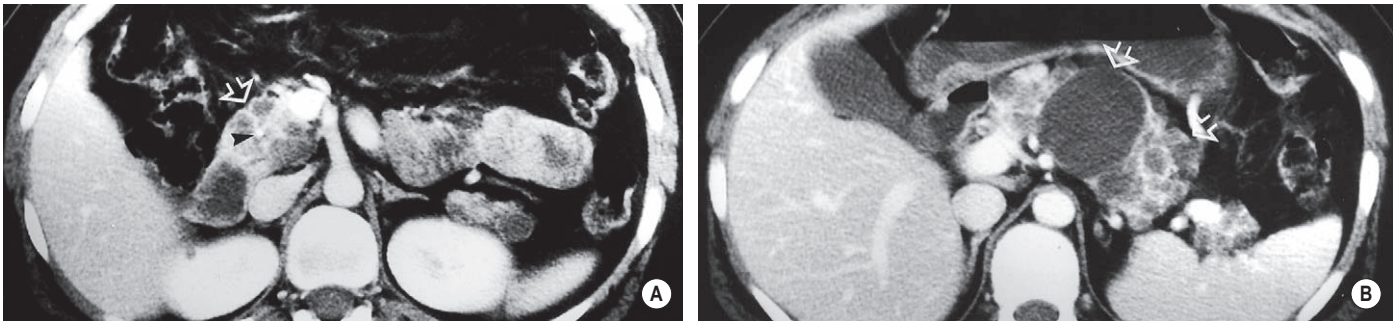
OSLER–WEBER–RENDU DISEASE

Definition A vascular disorder characterized by telangiectasia of the skin, mucous membranes, GI and urinary tracts, liver and pancreas

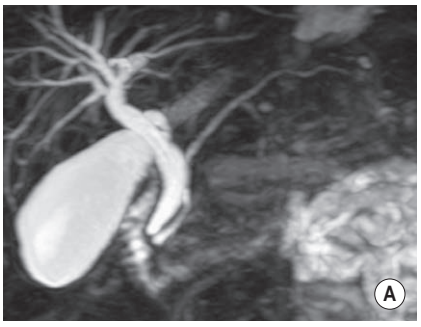
Angiography Dilated pancreatic arteries supplying a racemose collection of vessels with early draining veins

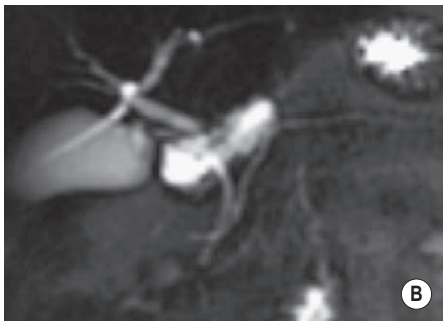


(A,B) Cystic fibrosis. CT through the level of the body and tail of the pancreas (arrows) shows fatty replacement of the gland.*

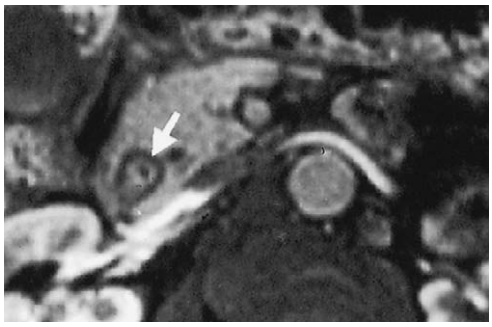


von Hippel–Lindau disease. (A) CT through the level of the head of the pancreas (open arrow) shows multiple small cysts and a central area of calcification (arrowhead) sited in a small serous cystadenoma. (B) Scan at the level of the body and tail of the pancreas (arrows) shows multiple cysts of different sizes. These were simple cysts, as distinct from the serous cystic neoplasm in the head of the pancreas.

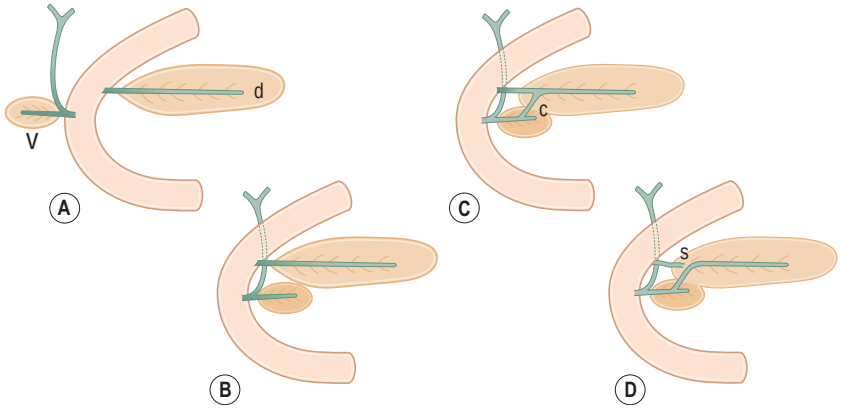




MRCP of normal pancreas and pancreas divisum. (A) Normal anatomy as demonstrated on MRCP. (B) Dorsal pancreatic duct drains into the minor papilla. The common bile duct drains separately into the major papilla.*



Annular pancreas. T1WI + Gad shows pancreatic tissue surrounding the second part of duodenum (arrow).†



Embryological development of the pancreas. (A) Dorsal segment (d) draining through the duct of Santorini and minor papilla. Ventral segment (v) developing in association with the bile duct and draining through the duct of Wirsung and major papilla. (B) The ventral segment has rotated with the bile duct to occupy its definitive position. This is the arrested embryological position of the adult pancreas divisum. (C) A wide communication (c) has developed between the dorsal and ventral ducts. (D) The terminal portion of the dorsal duct or duct of Santorini (s) becomes relatively smaller and may disappear completely. This is the normal adult arrangement.

ACUTE PANCREATITIS

Definition Acute inflammation of the pancreas ► it may be caused by the reflux of bile and pancreatic enzymes into the pancreatic parenchyma

- *Causes:* cholelithiasis (50%) ► idiopathic (10–30% – possibly related to congenital duct anomalies such as pancreas divisum) ► alcohol (20–25%) ► trauma ► surgery ► metabolic (hyperlipidaemia and hypercalcaemia) ► viral infection (mumps, cytomegalovirus and AIDS) ► drugs (steroids and thiazide diuretics)

Clinical presentation Epigastric pain ► nausea and vomiting ► raised serum amylase ► signs of haemorrhagic pancreatitis:

- *Cullen sign:* periumbilical bruising
- *Grey Turner sign:* flank bruising

Radiological features

XR Left-sided pleural effusion or atelectasis ► a gasless abdomen ► ascites ► intrapancreatic gas bubbles

- *'Colon cut off' sign:* a dilated transverse colon with an abrupt transition to a gasless descending colon
- *'Sentinel loop':* a localized segment of gas containing duodenum

Barium studies A widened duodenal sweep

- *'Frostberg inverted 3' sign:* this is due to segmental narrowing and fold thickening of the duodenum

US There is generalized (but less commonly focal) hypoechoic pancreatic enlargement ► ill-defined pancreatic margins ► peripancreatic fluid ► hepatic steatosis (if there is an associated high alcohol intake)

- US can exclude cholelithiasis as a cause, however it cannot reliably detect the presence of pancreatic necrosis

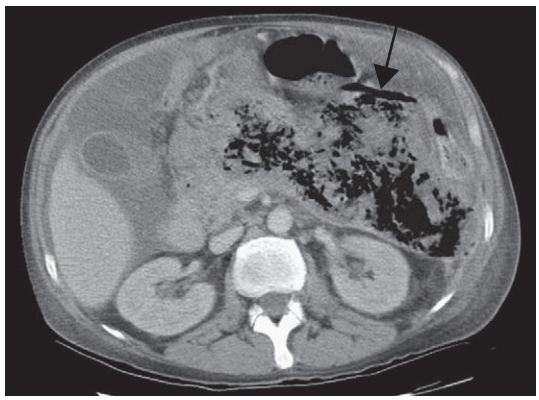
CECT

- *British Society of Gastroenterology Guidelines:* immediate CT is not indicated as the full extent of necrosis is only evident after 4 days (therefore the initial extent of necrosis may be underestimated) ► the contrast medium may also exacerbate any renal impairment
  - An immediate CT should only be performed if the extent of necrosis dictates the management or if the diagnosis is unclear
  - A follow-up CT is only required if there is a failure to improve or a clinical deterioration
- **Mild acute pancreatitis** (70–80%)
  - A normal or enlarged gland of uniform enhancement (± peripancreatic fat stranding and thickening of the fascial planes) ► cuffs of fluid may be seen around the adjacent vessels
- **Necrotizing acute pancreatitis** (a hallmark of severe acute pancreatitis)
  - There are non-enhancing pancreatic regions ► if this involves > 30% of the gland the mortality rate approaches 30%
  - *Infected necrosis (20–70%):* this is suggested if there are gas bubbles within any necrotic tissue (this can also be caused by a fistula to the GI tract) ► it is a major determinant of morbidity and mortality

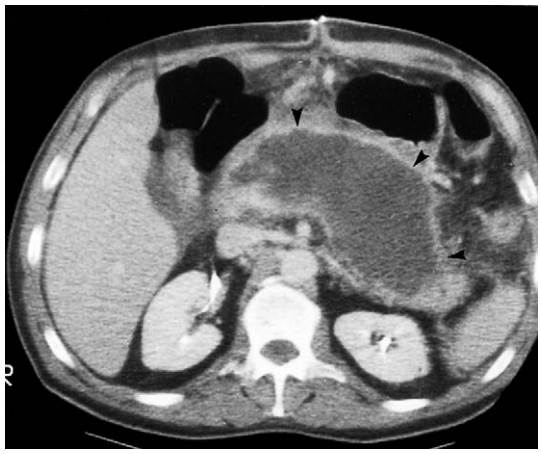
Pearls

- **Interstitial edematous pancreatitis (IEP)**
  - Homogeneous or slightly heterogeneous pancreatic enhancement
- **Acute necrotizing pancreatitis (sterile or infected)**
  - Parenchymal necrosis alone – non-enhancing pancreatic areas
  - Peripancreatic necrosis alone – commonly within retroperitoneum/lesser sac
  - Combined type (1 and 2) – the most common type
- **Pancreatic and peripancreatic collections (sterile or infected)**
  - Without necrosis:
    - *Acute peripancreatic fluid collections (APFC):* presents within 4 weeks and usually resorbed spontaneously without infection ► no non-liquefied components ► usually adjacent to the pancreas ► no discernable wall
    - *Pseudocyst:* after 4 weeks, an APFC may transition to a pseudocyst with a well-defined enhancing fibrous wall (containing no non-liquefied components) ► these rarely become infected
  - With necrosis:
    - *Acute necrotic collection (ANC):* presents within 4 weeks ► liquefaction within 2–6 weeks ► any collection replacing pancreatic tissue within 4 weeks is an ANC and not an APFC/pseudocyst
    - *Walled-off necrosis (WON):* after 4 weeks an ANC may transition to WON with a thickened non-epithelialized wall
- **Treatment**
  - *IEP/AFPC/pseudocyst:* usually self-limiting and spontaneously resolves
  - *Necrotizing pancreatitis:* if clinical status allows, supportive treatment for 2 weeks followed by surgical/radiological drainage as required
  - *Sterile pancreatic necrosis:* CT monitoring every 7–10 days to exclude complication or infection ► percutaneous drainage and supportive measures as required
  - *Infected pancreatic necrosis:* percutaneous drainage/surgical debridement

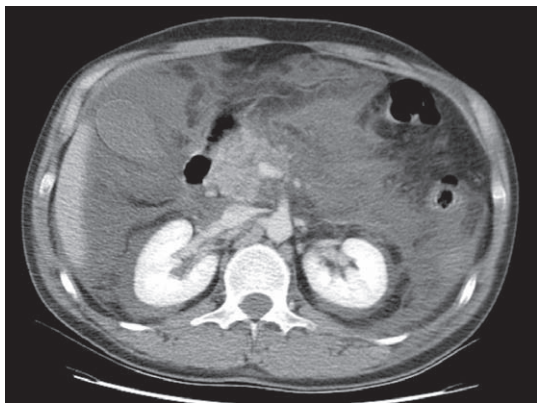
Mild acute pancreatitis. (A) Mild swelling of the gland that enhances uniformly but has indistinct margins because of peripancreatic oedema. There is inflammatory tissue around the coeliac axis (arrow). (B) Image at the level of the pancreatic head showing infiltration of the peripancreatic fat and fluid anterior to Gerota's fascia (arrows). (C) Different patient. Peripancreatic fluid with normally enhancing gland.*



Infected pancreatic necrosis. CECT shows non-enhancement of pancreatic tissue with secondary gas formation (arrow).



Pancreatic abscess. Dynamic CT shows a fluid collection with a contrast-enhancing capsule (arrowheads). Percutaneous aspiration showed the fluid to be pus. The abscess was drained successfully by percutaneous catheter.



Acute necrotizing pancreatitis. CECT shows enlargement of the pancreatic head, ascites and perirenal fluid collections. There is a largely necrotic, non-enhancing body and tail of the pancreas.*

ACUTE PANCREATITIS

Balthazar CT severity index (CTSI)	
CT features	**Score**
Grade	
Normal gland	0
Focal/diffuse enlargement	1
Peripancreatic inflammatory change	2
Single pancreatic fluid collection	3
Two or more fluid collections or abscess	4
Necrosis	
None	0
< 30%	2
30–50%	4
> 50%	6

CTSI vs morbidity and mortality		
Score	**Morbidity (%)**	**Mortality (%)**
0–3	8	3
4–6	35	6
7–10	92	17

PEARLS

Vascular involvement

- Intrapancreatic and peripancreatic arteries and veins may be eroded or thrombosed by pancreatic enzymes ► this may lead to pseudoaneurysm formation ► this can manifest as an episode of acute haemorrhage (due to vessel erosion, rupture of oesophageal, gastric, or mesenteric varices, or arterial pseudoaneurysm leakage)
- This requires CECT (± angiography and vascular embolization) ► ideally all cases should be assessed with a post-contrast pancreatic protocol CT to enable vascular assessment

GI involvement

- Direct extension of the inflammatory process can result in oedema, necrosis, or perforation of the stomach or duodenal wall
- Bowel involvement can occur due to pancreatic enzymes permeating through the mesenteric attachments (or secondary to vascular complications)

Bile duct involvement

- Oedema within the pancreatic head can cause a transient CBD obstruction
- Severe or chronic pancreatitis can cause a persistent CBD obstruction or stenosis

CHRONIC PANCREATITIS

DEFINITION

Irreversible pancreatic inflammation with an increased risk of pancreatic carcinoma

CLINICAL PRESENTATION

Chronic abdominal pain ► loss of exocrine and endocrine function (e.g. steatorrhoea and diabetes) ► weight loss

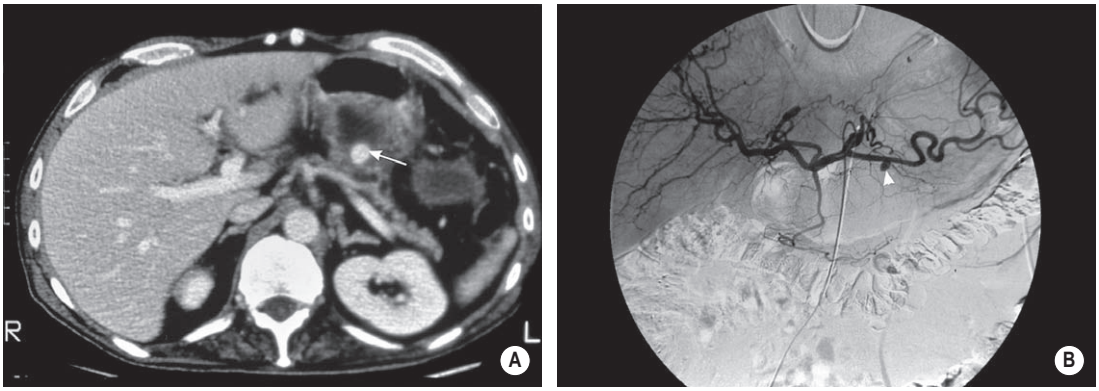
- *Causes:* idiopathic ► alcohol abuse ► hyperparathyroidism ► hyperlidaemia ► hereditary ► following multiple attacks of acute pancreatitis

RADIOLOGICAL FEATURES

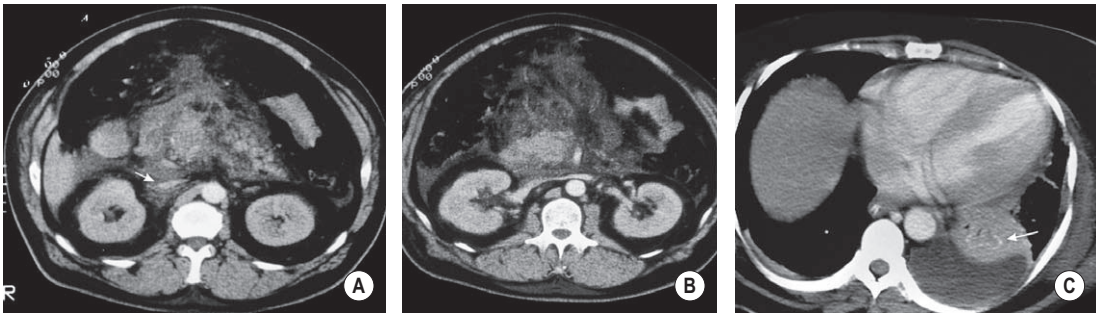
US/CT A heterogeneous pancreatic echotexture or attenuation ► parenchymal calcification (which may be seen on AXR) ► a dilated (>3mm) pancreatic duct (± CBD dilatation) ► atrophy or general pancreatic enlargement (focal enlargement can be mistaken for a pancreatic carcinoma) ► splenic, mesenteric or portal vein thrombosis ► arterial stenosis or occlusion ► arterial pseudoaneurysm formation

ERCP/MRCP Dilatation or multifocal stenoses of the main pancreatic duct and its lateral side branches ► intraductal filling defects (representing protein plugs or calcification) ► narrowing of the intrapancreatic CBD

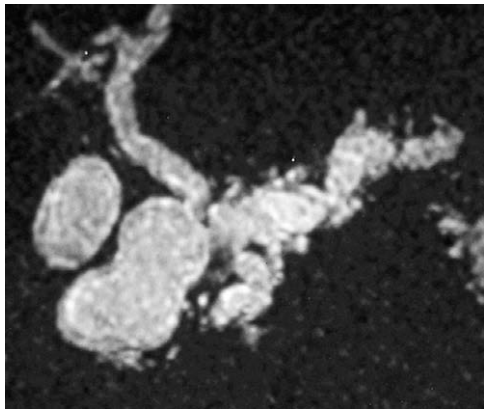
- *ERCP complications:* acute pancreatitis (in up to 10% of cases) ► haemorrhage ► cholangitis



Pseudoaneurysm formation. (A) CECT shows high-density contrast collection (arrow) within a small retrogastric pseudocyst. (B) Coeliac axis angiogram confirms the presence of a pseudoaneurysm (arrowhead). This was embolized successfully.



Moderately severe acute pancreatitis. (A) Inflammatory fluid is seen surrounding the inferior vena cava (arrow). (B) Extensive inflammatory changes involve the mesentery. (C) There is a left basal pleural effusion and collapse in the left lower lobe (arrow).*



Chronic pancreatitis. MR pancreatogram delineates a very abnormal main pancreatic duct with dilated side branches.

Chronic pancreatitis. Axial contrast-enhanced MDCT image shows diffuse calcifications in the pancreas with a dilated main pancreatic duct (arrows) measuring 9mm and dilated side branches.©20

PANCREATIC DUCTAL ADENOCARCINOMA

DEFINITION

- An aggressive pancreatic malignancy arising from the ductal epithelium and causing a local desmoplastic response with a propensity to constrict or obstruct adjacent ducts or vessels ► there is early involvement of the adjacent structures by perivascular, perineural and lymphatic spread
- Only 10% of tumours are resectable at diagnosis with an overall 5-year survival rate of only 2–3%
  - *At presentation:* liver metastases (50%) ► lymph node involvement (40%) ► peritoneal deposits (35%)

CLINICAL PRESENTATION

- Weight loss ► anorexia ► abdominal pain (due to invasion of the coeliac plexus) ► obstructive jaundice
- It is commonly seen within the 7^{th} to 8^{th} decades (M>F) ► smoking is a significant risk factor

RADIOLOGICAL FEATURES

- Approximately 70% of adenocarcinomas arise within the head, neck, or uncinate process – the remainder arise within the body or tail ► small masses may only be detectable by virtue of their differing imaging characteristics from normal tissue

US This differentiates obstructive from non-obstructive causes of jaundice ► a pancreatic tumour is a hypoechoic mass relative to the normal pancreas

Barium studies A widened duodenal loop ± mucosal irregularity (with spiculation and fold nodularity) ► a localized duodenal stricture or a double contour to the medial wall of the duodenal loop ► a 'reversed 3' sign of Frostberg

Cholangiography A pancreatic carcinoma produces a tight stricture that is often shouldered – this may have a blunt cut-off that is straight or convex upward or downward

CT Thin slices are used during the arterial phase and thicker slices during the portal phase acquisitions ► water is preferred to a positive oral contrast agent as the enhancement of the stomach and duodenal wall is more readily appreciated (allowing for the assessment of any invasion)

- Assessment with dual phase imaging:
  - *'Pancreatic' phase (late arterial enhancement):* as the tumour is hypovascular it will appear as a poorly enhancing focal area within densely enhancing normal pancreatic tissue
  - *Portal phase imaging:* this assesses for any vascular involvement and metastatic disease
- There is mass effect with alteration of the pancreatic contour ► the local desmoplastic response may mimic the appearances of a focal pancreatitis ► calcification is rare
- *'Double duct' sign:* this follows adjacent stricture formation within both the common bile and pancreatic ducts and is highly suggestive of a pancreatic carcinoma (less commonly a periampullary carcinoma) ► it leads to bile duct obstruction, upstream main pancreatic duct dilatation and pancreatic atrophy ► uncinate masses are often large before bile duct obstruction occurs (due to their location) ► chronic pancreatitis may produce this sign but it will also demonstrate other distinguishing features (e.g. pancreatic calcification)
- *Courvoisier's sign:* an enlarged, non-tender and thin-walled gallbladder secondary to distal biliary obstruction (cf. cholecystitis with an enlarged, tender, thick-walled gallbladder)
- There is often retropancreatic extension with obliteration of the fat planes around the coeliac axis and SMA (indicating unresectable disease)

MRI This offers no significant advantage over CT ► T1WI (FS): tumour is of lower SI than the normal pancreas ► T2WI: there is often high SI (but this can be variable) ► T1WI + Gad: poor enhancement

ERCP This directly visualizes the duodenum and ampulla of Vater ► it also allows for cytological sampling and access for stent insertion

EUS and laparoscopic US This may be a helpful supplement in patients with potentially resectable disease ► although it can demonstrate a hypoechoic mass, there is poor visualization of the coeliac axis and splenic artery

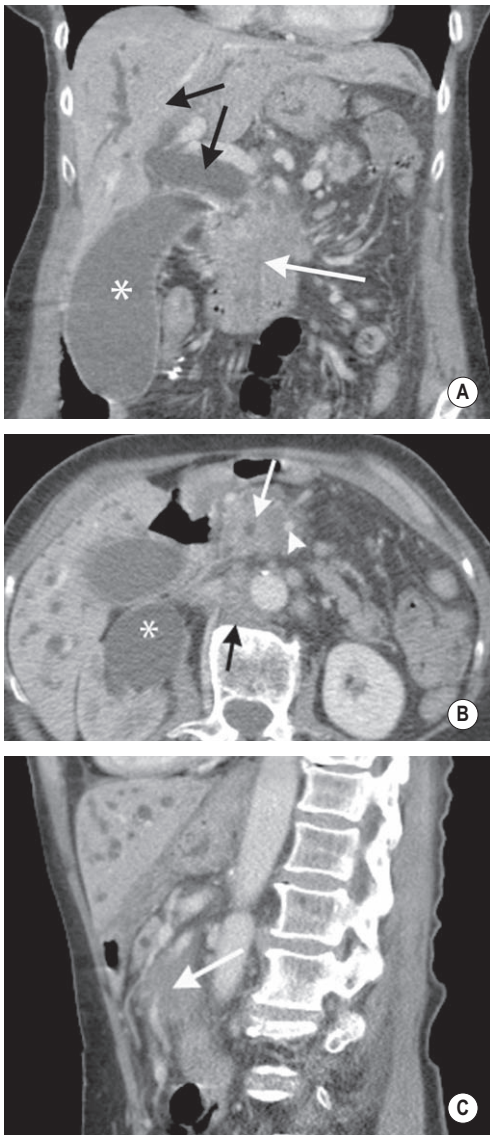
FDG PET This has a limited role: it cannot distinguish between a malignant or inflammatory mass

- However it demonstrates high accuracy in the detection of local recurrence

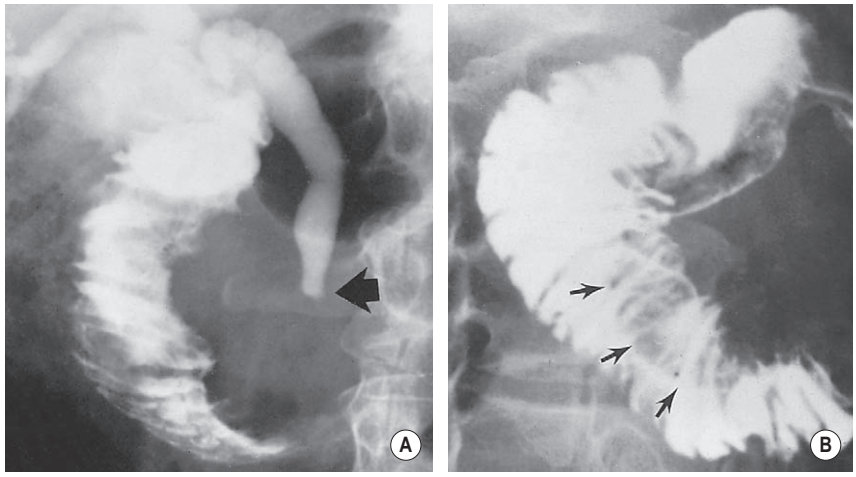
PEARLS

Assessment of resectability (CT/MRI):

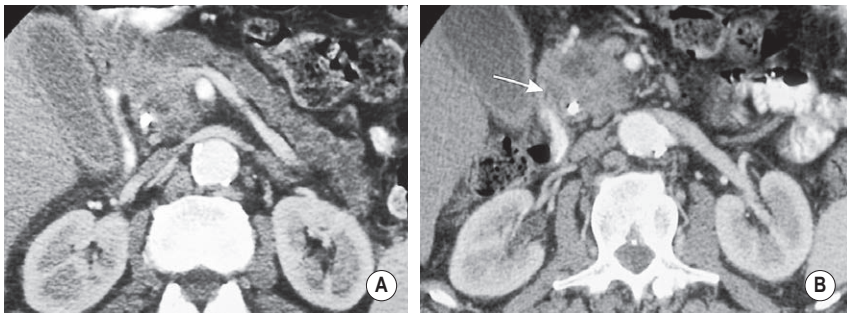
- In the absence of distant metastatic disease, perivascular invasion or vascular encasement are the most important criteria for unresectability ► tumours of the body and tail tend to be unresectable as these tend to have metastases at presentation
  - *CT findings indicating perivascular invasion*: soft tissue infiltration obscuring the vessel margin ► a vessel calibre change or contour deformity
    - Complete arterial occlusion may result in splenic infarction
    - Tumours within the pancreatic head and uncinate process tend to affect the SMA ► tumours within the body and tail tend to affect the coeliac, hepatic or splenic arteries



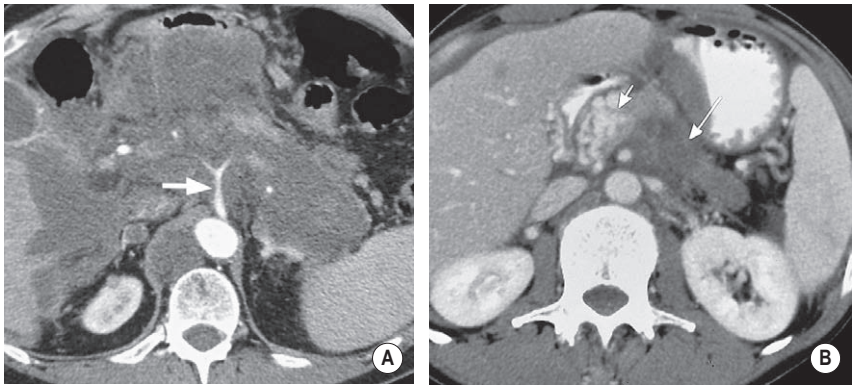
Pancreatic carcinoma. (A) Coronal CT demonstrating a mass within the pancreatic head (white arrow) with a significant associated desmoplastic response. There is associated common biliary and intrahepatic bile duct obstruction (black arrows), together with a distended thin-walled gallbladder (asterisk) (Courvoisier's sign). (B) Axial image demonstrating the pancreatic head mass within which can be seen the common bile duct (arrow). The mass is totally encasing the superior mesenteric vessels (arrowhead), making this unresectable disease. In addition, there is retroperitoneal spread that is encasing approximately 50% of the aortic circumference (black arrow). The right hydronephrosis (asterisk) is secondary to an obstructive tumour deposit within the pelvis. (C) Sagittal reformat demonstrating the tumour mass encasing the SMA (arrow).



(A) Enlarged duodenal loop with a 'reversed 3' sign of Frostberg. Earlier PTC shows the characteristic 'gloved finger' obstruction of the intrapancreatic common bile duct pathognomonic of carcinoma of the pancreatic head. (B) Barium meal. A double contour (arrows) of the duodenal loop. Carcinoma of the pancreatic head.



Pancreatic carcinoma. (A) Atrophy of the body and tail is seen, with a markedly dilated pancreatic duct. (B) The pancreatic head mass involves the duodenal wall (arrow).



(A) Pancreatic carcinoma and adjacent adenopathy encasing the coeliac axis (arrow). (B) Venous occlusion. A tumour in the pancreatic body (long arrow) has occluded the splenic and portal veins, resulting in the development of multiple venous collaterals (small arrow). Note the presence of splenomegaly and a hepatic metastasis.

PANCREATIC DUCTAL ADENOCARCINOMA

PEARLS

- *Indirect CT findings of venous involvement* (often involving the superior mesenteric and splenic veins at the portal venous confluence): dilatation of small peripancreatic veins (± multiple venous collaterals) ► a 'tear drop' shape of the SMV
- *Unresectable disease*: circumferential vascular encasement (particularly if the tumour exceeds 50% of the vessel circumference)
- SMV encasement (but a normal SMA) will not itself preclude resection

- *Invasion of adjacent structures (e.g. stomach, duodenum):* this will be shown by interruption of their normally enhancing wall ► duodenal involvement does not necessarily preclude curative surgery as the duodenum is removed as part of the procedure
- *There is early metastatic spread to the lymph nodes*: peripancreatic followed by coeliac, common hepatic, mesenteric and then para-aortic ► nodes may be involved without enlargement (nodes measuring > 1cm in short axis are suspicious for metastatic involvement)
- *Peritoneal spread:* this is commonly seen but the lesions are typically small and difficult to detect ► peritoneal involvement may be inferred if there is ascites
- *Distant metastases*: liver > lymph nodes > peritoneum > lung

Tumour markers (CA 19-9, CA 242, CEA) These are associated with pancreatic cancer but are not currently sensitive or specific enough for screening or the differentiation of benign from malignant pancreatic masses

Diagnosis This can be achieved with FNA biopsy ► this should be possibly avoided if the tumour is potentially resectable (due to the risk of cutaneous seeding)

Treatment Whipple's procedure: a radical pancreaticoduodenectomy with gallbladder removal ± distal gastric resection (a jejunal loop creates a gastrojejunal, choledochojejunal and pancreatojejunal anastamosis)

- This is associated with significant morbidity and mortality
- A radiologically guided coeliac axis block can help with pain relief

CYSTIC PANCREATIC TUMOURS

DEFINITION

- The commonest cystic mass within the pancreas is a pseudocyst (demonstrating a high amylase level following FNA) ► if there is a low amylase level then a cystic tumour has to be considered:

Intraductal papillary mucinous neoplasm (IPMN)

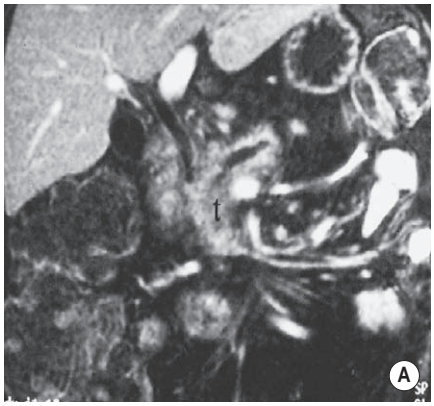
This is a rare cystic low-grade pancreatic tumour arising from the epithelial lining of the pancreatic ducts ► excessive mucin secretion results in duct dilatation and obstruction (it can involve the main or side ducts)

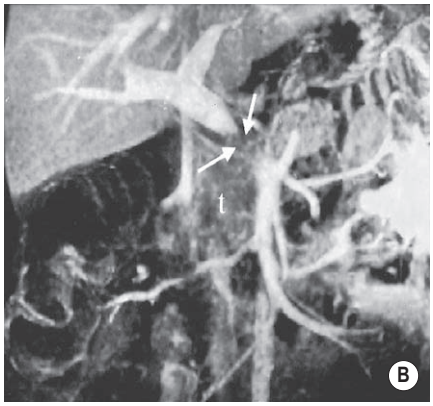
- *Location:* pancreatic head (58%) > body (23%) > tail (7%)
  - *Main duct IPMN:* high malignancy risk (invasive features in 50%)
    - – Surgical removal usually advocated
  - *Branch duct IPMN:* lower malignancy risk (especially if < 3cm)
    - – Close follow-up advocated if < 3cm and no signs of malignancy
  - Predictors of malignancy: main pancreatic duct > 9mm ► mural enhancing nodules ► signs of invasion ► thick septa ► irregular wall

MRCP/MDCT/EUS/ERCP These can potentially detect a communication between the IPMN and pancreatic duct (no such communication exists with a mucinous cystic neoplasm)

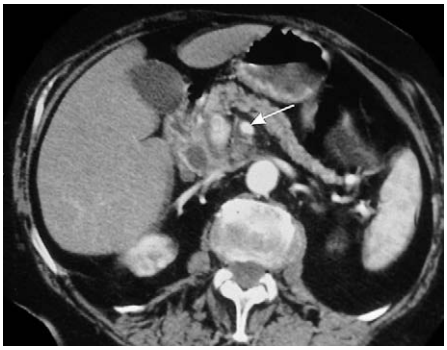
Serous cystadenoma vs mucinous cystic tumour

	Mucinous cystic neoplasm (malignant potential)	Serous cystadenoma (benign potential)
Frequency	More common	Less common
Location	Body or tail of the pancreas	Head of the pancreas
Age	Younger patients	Older patients
Morphology	Fewer larger cysts (<6 cysts, >2cm in diameter)	Numerous tiny cysts (>6 cysts, <2cm in diameter) ► they may appear solid on CT if the cysts cannot be resolved
Central scar	Absent	Present
Calcification	Amorphous peripheral mural calcification	Central stellate calcification ('sunburst')
Vascularity	Hypovascular	Hypervascular

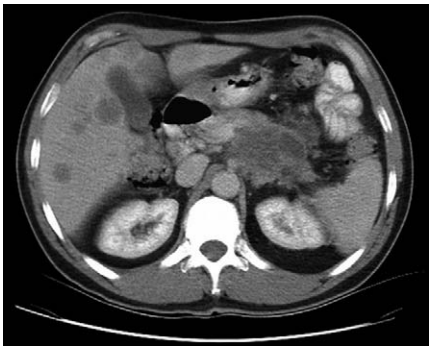




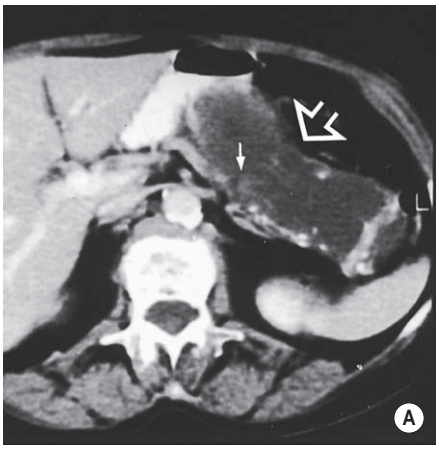
Unresectable carcinoma of the pancreas. (A) Coronal T1WI + Gad shows the ducts are obstructed by an ill-defined tumour (t), which is of slightly lower SI than adjacent pancreas ► maximum intensity projection (B) shows the lower end of the portal vein to be encircled (arrows) by extension of the tumour (t) from the head of the pancreas.

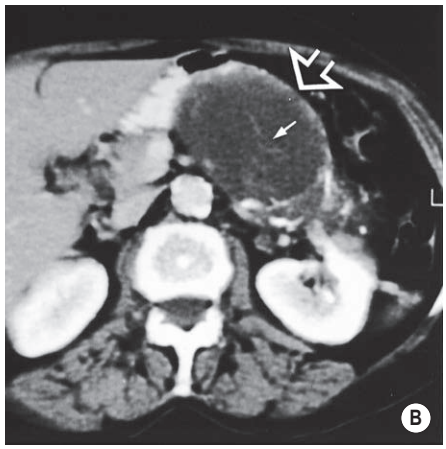


Unresectable pancreatic carcinoma. The uncinate process tumour encircles more than 50% of the circumference of the superior mesenteric artery (arrow).



Pancreatic carcinoma with liver metastases. Portal venous phase CT shows poorly enhancing liver deposits secondary to a primary mass in the pancreatic body.





Mucinous cystadenocarcinoma. (A,B) Contiguous CT images through the pancreas show a cystic mass (open arrow) replacing most of the body and tail of the gland. Areas of dystrophic calcification are noted within the wall of the cystic mass and small papillary excrescences and septa are also seen (small white arrows).*

Benign serous cystadenoma. The tumour at the junction of the pancreatic body and tail shows the typical appearance of numerous small cysts.*

ENDOCRINE PANCREATIC TUMOURS AND ISLET CELL TUMOURS

DEFINITION

Functioning tumours

- Peptide hormone production produces a characteristic clinical syndrome

Insulinoma

- **Definition:** the commonest islet cell tumour (accounting for 50% of the total) presenting with hypoglycaemic episodes ► the diagnosis can be confirmed biochemically
- **Location:** there is no predilection for any particular part of the pancreas
- **Characteristics:** the tumour is usually solitary and small (<2cm) ► 90% are benign
  - *'10% rule':* 10% are associated with multiple endocrine neoplasia syndrome (MEN) type 1 (10%) ► 10% are multiple ► 10% are malignant

Gastrinoma

- **Definition:** the 2nd commonest islet cell tumour presenting with the Zollinger–Ellison syndrome (gastric hyperacidity with recurrent gastric and duodenal ulceration)
- **Location:** these can be ectopic (e.g. within the duodenal wall, stomach, or omentum)
  - *They are frequently found within the 'gastrinoma triangle':* the porta hepatis forms the triangle apex with the 2nd and 3rd parts of the duodenum forming the base
- **Characteristics:** they are often multiple, with a mean size of 3.5cm ► 60% are malignant
  - ⅓ are associated with MEN type 1

Glucagonoma

- **Definition:** this presents with diarrhoea, diabetes, necrolytic erythema migrans and glossitis (secondary to excessive glucagon secretion)
- **Location:** tumours are predominantly located within the pancreatic body and tail
- **Characteristics:** the average tumour size is 4–7cm ► 60% are malignant

VIPoma

- **Definition:** this secretes vasoactive intestinal polypeptide leading to the WDHA syndrome (**w**atery **d**iarrhoea, **h**ypokalaemia, **a**chlorydria) ► malignant transformation is seen in 60% of cases
- **Location:** tumours are predominantly located within the pancreatic body and tail ► 10% are ectopic (and found within the sympathetic chain and adrenal medulla)
- **Characteristics:** the average tumour size is 5–10cm ► most tumours are benign, but 50% of intrapancreatic tumours are malignant

Somatostatinoma

- **Definition:** this presents with hyperglycaemia, gallstones and steatorrhoea (secondary to excessive somatostatin secretion)
- **Location**: tumours are located within the pancreatic head or duodenum
- **Characteristics:** the average tumour size is >4cm ► >50% of tumours undergo malignant transformation

Non-functioning tumours

- **Definition:** these are the 3rd commonest islet cell tumour
- **Location:** they are commonly found within the pancreatic head
- **Characteristics:** they can be large at presentation (> 5cm), causing symptoms by mass effect (e.g. jaundice) ► they are nearly always malignant
- **Clinical presentation:** this is similar to a pancreatic adenocarcinoma – however there is a better prognosis as they can often be curatively resected or successfully treated with chemotherapy

RADIOLOGICAL FEATURES

US This detects > 60% of solitary islet cell tumours ► they appear as a well-defined lesion of low reflectivity compared with the adjacent pancreas

Intraoperative US This is carried out if the preoperative location was unsuccessful ► it can visualize tumours as small as 3mm (endoscopic and intraductal US is also available)

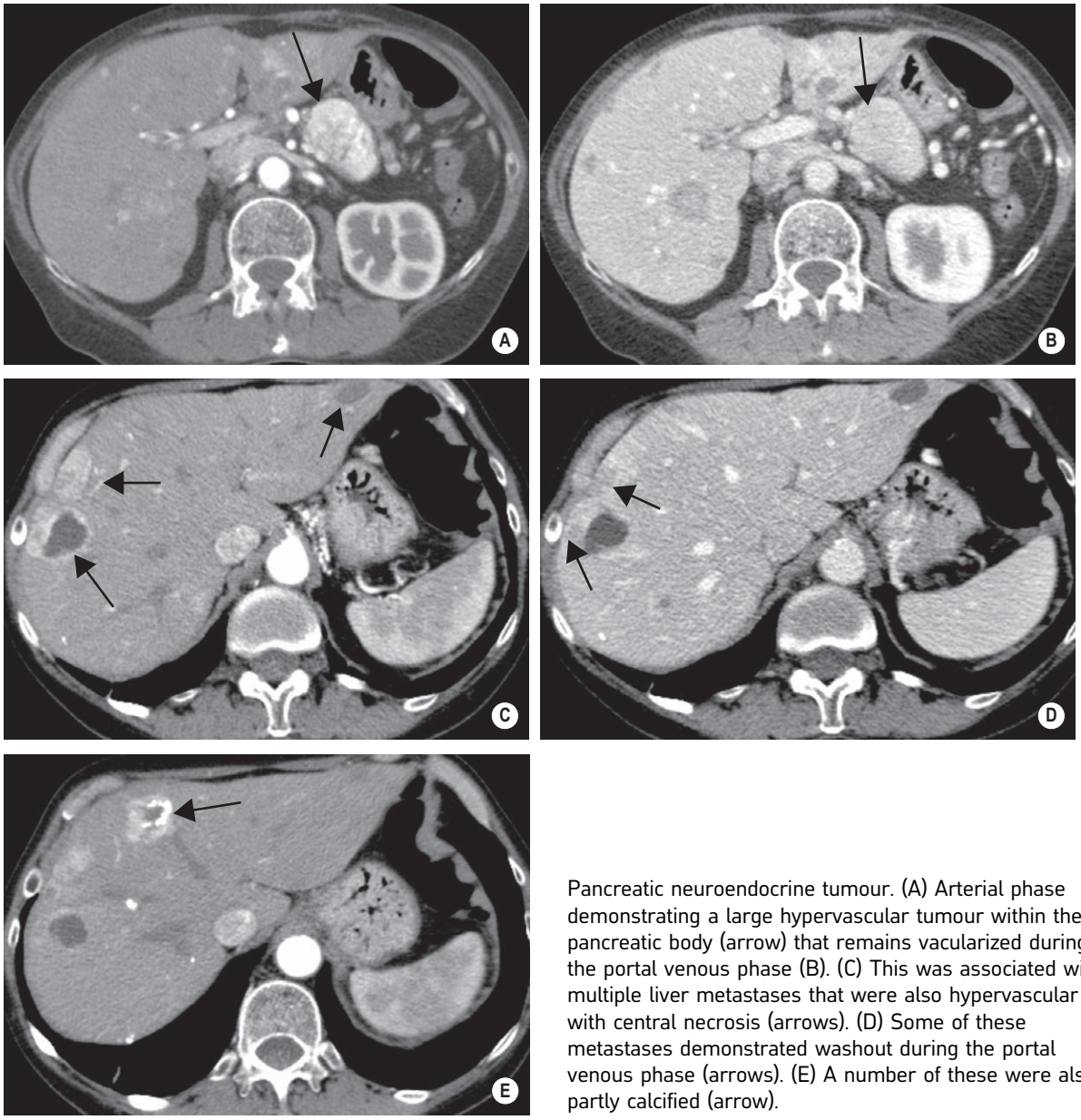
CT A tumour demonstrates early but transient enhancement (as it is hypervascular) ► there may be better depiction on portal phase imaging (there may be ring enhancement with an insulinoma)

- Radiological features suggesting a non-functioning endocrine tumour as opposed to an adenocarcinoma:
  - *Calcification (22%):* this is rarely seen in an adenocarcinoma
  - *Contrast enhancement:* this is not a feature of an adenocarcinoma

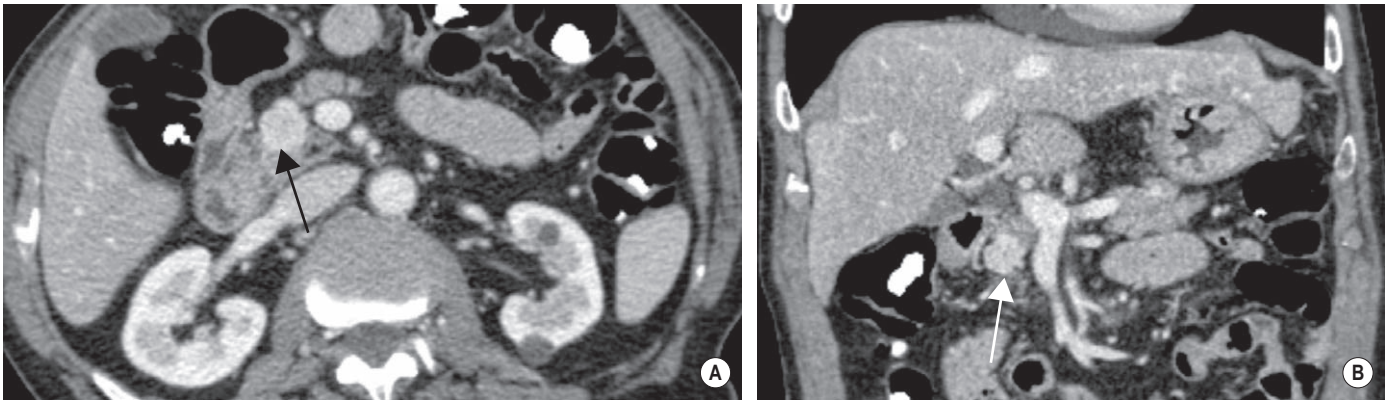
MRI T1WI (FS): low SI ► T2WI: high SI ► T1WI + Gad: solid or ring enhancement

Scintigraphy ^{111}In-pentetreotide and ^{123}I-MIBG can localize the appropriate cell type ► PET can be utilized (using tracers other than traditional FDG)

Venous sampling This may localize a functioning tumour ► the hepatic vein is sampled after selective pancreatic arterial injection of a secretagogue (calcium for an insulinoma and secretin for a gastrinoma)



Pancreatic neuroendocrine tumour. (A) Arterial phase demonstrating a large hypervascular tumour within the pancreatic body (arrow) that remains vacularized during the portal venous phase (B). (C) This was associated with multiple liver metastases that were also hypervascular with central necrosis (arrows). (D) Some of these metastases demonstrated washout during the portal venous phase (arrows). (E) A number of these were also partly calcified (arrow).



Pancreatic neuroendocrine tumour. (A) A small hypervascular tumour is seen within the pancreatic head (arrow), which is also seen on the coronal view (B).

TRAUMA

DEFINITION

- Trauma may result in pancreatic contusion, laceration or a complete transection ► it is uncommon
- *Mechanism:* a severe direct impact or a forceful deceleration injury with midline compression of the pancreas against the vertebral column
  - Pancreatic injuries are often associated with other visceral injuries
- Blunt pancreatic injuries:
  - *Without ductal leakage:* these usually resolve spontaneously
  - *With ductal leakage:* post-traumatic pancreatitis may occur ► disruption of the main pancreatic duct is an important indicator of severity
  - *Other complications:* abscess, fistula, or pseudocyst formation

RADIOLOGICAL FEATURES

US Peripancreatic fluid ► discontinuity of the normal pancreatic contour

CT This is the best investigation

- A pancreatic fracture line (± separation of the fragments) ► haematoma formation ► focal enlargement of the pancreas ► fluid lying between the splenic vein and pancreas ► increased peripancreatic fat attenuation ► thickening of the anterior renal fascia ► fluid within the lesser sac

ERCP/MRCP This is performed if a ductal injury is suspected

PANCREATIC TRANSPLANT IMAGING

- This is increasingly used for the treatment of diabetes (± a simultaneous renal transplant)

Imaging can detect rejection

US (acute rejection) Patchy or diffuse areas of decreased parenchymal echogenicity ► an enlarged graft

US (chronic rejection) Increased echogenicity ► a reduced graft size

Scintigraphy ^{99m}Tc-DTPA blood pool imaging: there is reduced graft perfusion

MRI This is the most sensitive technique ► T1WI: reduced SI (similar to skeletal muscle) ► T2WI: increased SI (similar to fluid)

Imaging can detect other complications

Transplant pancreatitis and associated perigraft fluid collections These are not uncommon and can be treated with percutaenous catheter drainage

Disruption of the cystoduodenostomy and an anastomotic leak This can be demonstrated with a CT cystogram

Other complications Abscess formation ► haemorrhage ► ischaemia ► graft-vessel thrombosis

INTERVENTIONAL RADIOLOGY IN THE PANCREAS

BIOPSY OF A PANCREATIC LESION

US-guided biopsy Using an anterior approach

CT-guided biopsy Using an anterior, posterior, or even a lateral approach

- It is usually necessary to pass the needle through normal abdominal tissue – most structures (except the spleen) can be traversed with a 20 or 22G needle without significant morbidity
  - *FNA biopsy:* this provides a cytological aspirate
  - *Cutting needles (18–20G):* these provide a core of tissue for histology

Complications Pancreatitis (the commonest complication – 3%) ► a vasovagal reaction ► severe haemorrhage ► needle-track seeding (rarely seen)

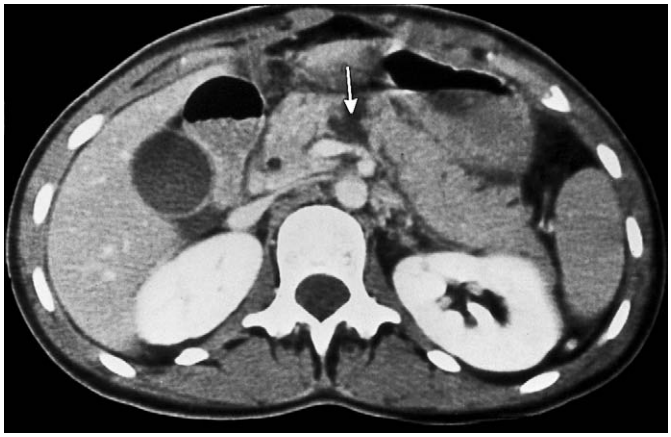
PERCUTANEOUS DRAINAGE OF A PANCREATIC FLUID COLLECTION

- This usually follows acute pancreatitis with 2 main indications:
  - *To assess whether a fluid collection is infected:* a few millilitres are aspirated for microbiology
  - *Drainage of a known infected collection:* the best results are obtained using large catheters (≥12F as most abscesses are viscous) ► drainage of separate collections is achieved with additional catheters
  - Drainage of a pancreatic collection following enzymatic destruction of pancreatic tissue is usually delayed by 2 weeks to allow the collection to sufficiently liquefy ► other peripancreatic collections can be drained immediately

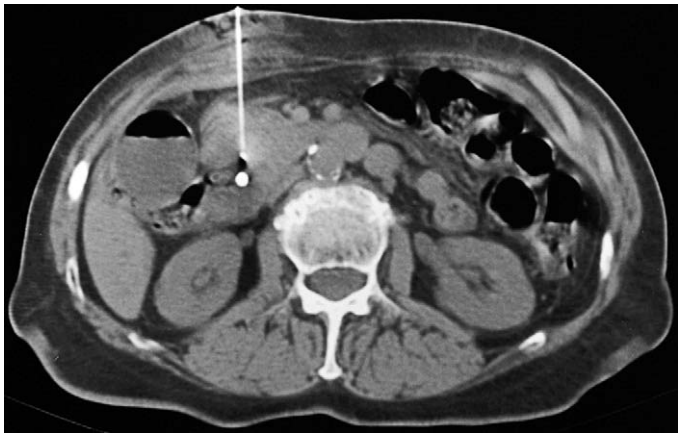
OTHER INTERVENTIONAL RADIOLOGICAL PROCEDURES

Percutaneous access into dilated pancreatic ducts For either balloon dilation or stenting of benign pancreatic duct strictures

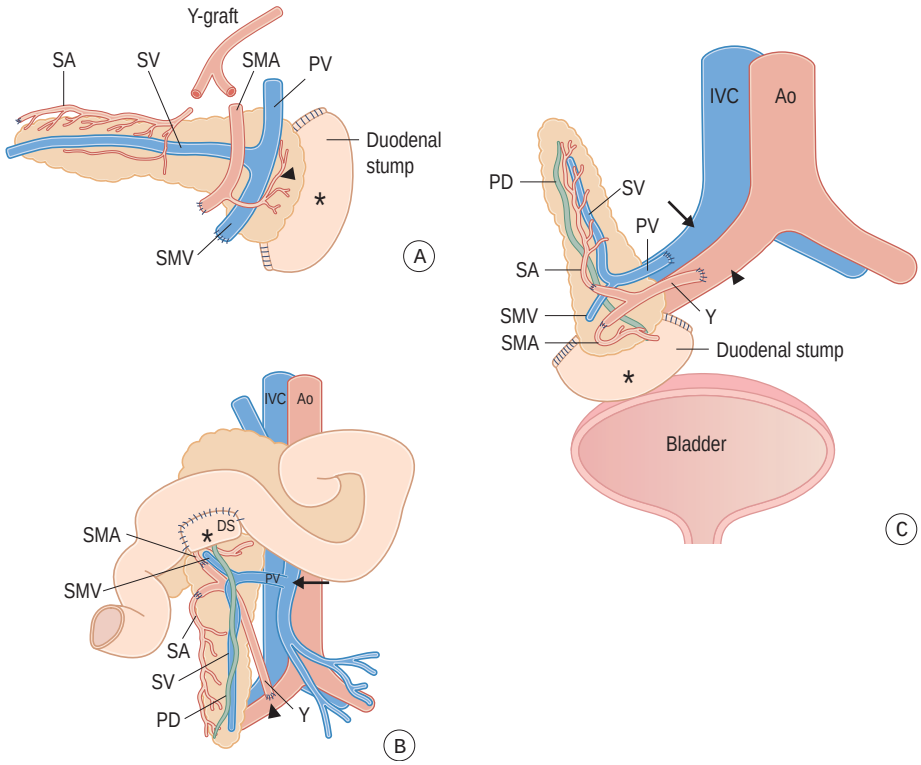
Percutaneous cystgastrostomy Creation of a communication between a pancreatic pseudocyst and the stomach (with the percutaneous insertion of a drainage catheter between the pseudocyst and stomach)



Pancreatic trauma. Following direct blunt trauma to the abdomen the pancreas is seen to be fractured (arrow). There was disruption of the main pancreatic duct.



CT-guided biopsy of the pancreas. A pancreatic mass is present and there is a plastic biliary stent in situ. Using an anterior approach, a 22-gauge needle has been inserted into the mass for biopsy. Fine-needle aspirate revealed adenocarcinoma.*



(A) Drawing of the donor pancreas (posterior view) demonstrates vascular reconstruction. The donor iliac Y-graft is attached to the donor splenic artery (SA) to supply the body of the pancreas, and to the donor superior mesenteric artery (SMA) to supply the head of the pancreas via the inferior pancreaticoduodenal artery (arrowhead). The donor portal vein (PV) functions as the main graft vein and drains the donor splenic vein (SV) and donor superior mesenteric vein (SMV). The donor duodenal stump (*) is harvested along with the pancreas. (B, C) Drawings (anterior view) illustrate the two types of pancreas transplants as seen through the overlying pancreas. (B) Portal venous and enteric exocrine drainage. The graft artery (Y) is attached to the common iliac artery (arrowhead) proximally ► the distal limbs are connected to the donor splenic artery (SA) and donor superior mesenteric artery (SMA). The graft vein (PV) is attached to the recipient superior mesenteric vein (arrow) for portal venous drainage. The donor splenic vein (SV) and donor superior mesenteric vein (SMV) are shown as well. Exocrine drainage is via the pancreatic duct (PD) to the duodenal stump (*), which is anastomosed to the jejunum. Ao = aorta, IVC = inferior vena cava. (C) Systemic venous and bladder exocrine drainage. The graft artery (Y) is attached to the common iliac artery (arrowhead) proximally ► the distal limbs are connected to the donor splenic artery (SA) and donor superior mesenteric artery (SMA). The graft vein (PV) is anastomosed to the recipient external iliac vein (arrow), providing systemic venous drainage of the donor splenic vein (SV) and donor superior mesenteric vein (SMV). Exocrine secretions are drained via the pancreatic duct (PD) to the duodenal stump (*), which is anastomosed to the bladder. Ao = aorta, IVC = inferior vena cava. ©21

3.9 SPLEEN

NORMAL VARIANTS AND CONGENITAL ANOMALIES

- **Splenunculus:** this represents ectopic splenic tissue of congenital origin ► they can be single or multiple and are usually found at the splenic hilum ► they have similar imaging appearances to the spleen
- **Wandering spleen:** this follows laxity of the suspensory ligament ► torsion may occur
- **Polysplenia:** the spleen is divided into 2–16 masses ► this is a congenital syndrome associated with situs ambiguous, as well as cardiovascular and visceral anomalies
- **Asplenia (right isomerism):** an absent spleen and multiple anomalies are seen within the abdomen and thorax (e.g. situs ambiguous with right sidedness)
- **Splenogonadal fusion:** congenital fusion of splenic tissue and the gonad (usually left sided) ► it usually affects males

SPLENIC INFECTION

- **Bacterial abscesses (staphylococci, streptococci or salmonella):** this presents with rim-enhancing collections
- **Mycobacterium tuberculosis:** the miliary form spreads by haematogenous dissemination ► there can also be splenic infarcts, peritoneal TB, and adenopathy
- **Fungal infection (candida, aspergillus and cryptococcus):** this occurs in immunosuppressed patients ► it presents with miliary, multifocal low-density lesions demonstrating central high density on CT ('bull's eye' lesions) ► there can be tiny calcifications present (2–5 mm)

SPLENOSIS

- Heterotopic autotransplantation of splenic tissue, which usually follows a traumatic rupture of the splenic capsule
  CT Poorly marginated soft tissue nodules
- *Possible features:* subcapsular or intrasplenic haematoma ► laceration ► infarction ► haemoperitoneum ► pseudoaneurysm formation

BENIGN MASS LESIONS

Splenic cyst

- *True (primary) cyst:* this has a cellular lining ► it can be congenital (F>M) or secondary to echinococcal infection
- *False (secondary) cyst:* this does not have a cellular lining ► it is usually post-traumatic and probably represents an evolved haematoma

Haemangioma

- This is the commonest primary benign neoplasm of the spleen ► it may be part of the Klippel–Trénaunay–Weber syndrome
  - Large haemangiomas may lead to splenic rupture and anaemia, thrombocytopenia and coagulopathy (Kasabach–Merritt syndrome)

Lymphangioma

- These are multiple thin-walled, well-defined cysts ► they can be capillary, cavernous or cystic (the commonest type within the spleen) ► they are often subcapsular and can be single or multiple ► they are usually asymptomatic
  CT They do not enhance

Hamartoma

- A rare benign lesion composed of an anomalous mixture of normal splenic elements ► they are usually single lesions
  US A hyperechoic (± a cystic component) lesion
  NECT An isodense or hypodense lesion
  MRI T1WI: intermediate SI ► T2WI: high SI ► T1WI + Gad: slow enhancement with late filling in

MALIGNANT MASS LESIONS

Lymphoma and leukaemia

- Splenic lymphoma is usually part of a generalized lymphoma (usually NHL (non-Hodgkin's lymphoma)) ► there is splenomegaly (± multifocal deposits) – see separate section

Splenic angiosarcoma

- This is very rare but is the commonest non-lymphoid primary malignant tumour of the spleen ► there is a very poor prognosis

Other primary tumours

- These are very rare (e.g. fibrosarcoma, leiomyosarcoma)

Metastatic disease

- This is rare and usually asymptomatic ► parenchymal lesions appear as hypodense nodules on CT
  - *Commonest primary sites*: breast ► lung ► colorectal ► ovary ► skin (melanoma)
    - There may be cystic metastases from the ovary, breast, endometrium or skin
  - Calcification is uncommon (but can be seen with mucinous primary adenocarcinomas)
  - Ovarian, gastrointestinal or pancreatic tumours can lead to peritoneal splenic disease

OTHER SPLENIC DISORDERS

Splenic infarcts

CT Low-density, wedge-shaped areas on CT – infarct of the whole spleen results in only rim enhancement of the capsule

- *Causes:* embolic disease ► arteritis ► sickle cell disease

Splenic sarcoidosis

CT Hypodense nodules ► splenomegaly ► adenopathy

Haemosiderosis

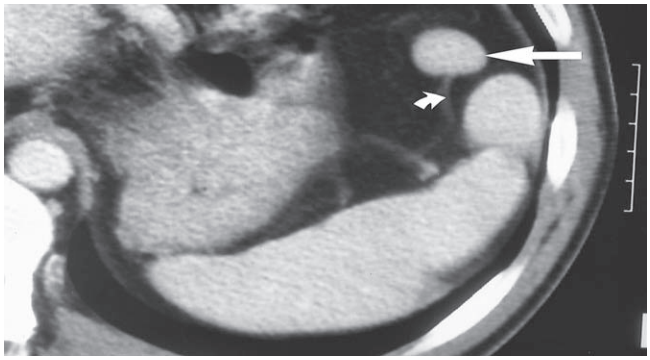
MRI T1WI and T2WI: there is low SI due to the iron deposition

Amyloidosis

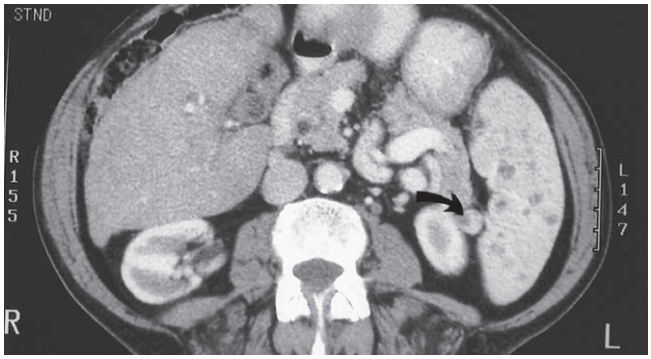
CT Low-density lesions within an enlarged spleen

Sickle cell anaemia

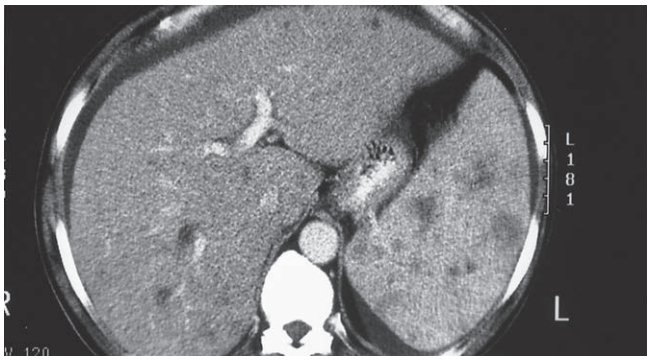
XR/CT With chronic disease there is a small calcified spleen (due to repeated splenic infarction)



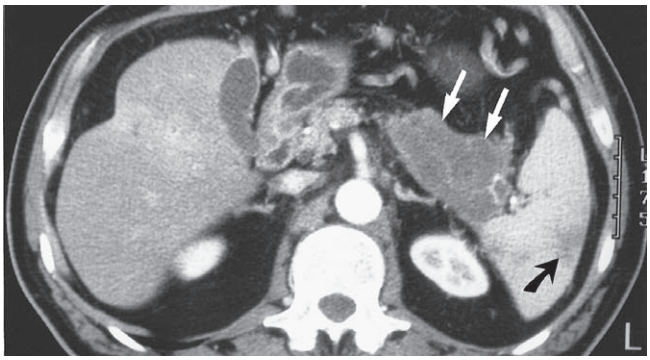
Accessory spleen. CECT shows a small accessory spleen (arrow) anterior to the spleen. Note the draining vein (curved arrow) from the accessory spleen can be seen to join the other veins draining the main spleen.*



Splenic candidiasis. CECT shows multiple tiny hypodense lesions in the spleen from candidal microabscess. A small accessory spleen (curved arrow) at the splenic hilum is also involved.*

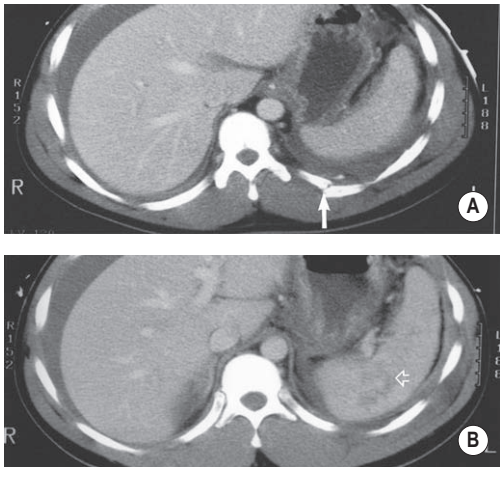


Lymphoma. CECT in a patient with NHL involving the liver and spleen. Multiple small poorly defined nodules can be seen in both the liver and spleen. A trace of ascites can also be seen anterior to the liver.*



Splenic infarct. CECT shows a tumour mass (white arrows) in the tail of the pancreas causing occlusion to the splenic vein and a splenic infarct (black arrow).*

Splenomegaly		
A huge spleen	**A moderately large spleen**	**A slightly large spleen**
	As for a huge spleen but also:	*As for a huge and moderately large spleen but also:*
• Chronic myeloid leukaemia • Myelofibrosis • Malaria • Gaucher's disease • Lymphoma	• Storage diseases • Haemolytic anaemias • Congestive causes (portal hyptertension, cirrhosis, cystic fibrosis, splenic vein obstruction) • Leukaemia	• Infections (hepatitis, malaria, EBV, TB, typhoid) • Sarcoidosis • Amyloidosis • Rheumatoid arthritis (Felty's syndrome) • SLE
©12		



Splenic injury. CECT in a man who fell from a ladder. (A)There is fracture of the left 10th rib (arrow) with perisplenic fluid and ascites. (B) Splenic laceration (open arrow) was also identified on CT.*

3.10 PERITONEUM, MESENTERY AND OMENTUM

ASCITES

DEFINITION

- >100ml of free fluid within the peritoneal cavity due to benign or malignant causes
- *Sequence of peritoneal fluid movement:* it initially collects around the liver ► it then flows to the pouch of Douglas ► it then flows symmetrically to both lateral paravesical spaces ► it finally ascends both paracolic gutters (due to negative intra-abdominal pressures during respiration)
  - It preferentially ascends the right paracolic gutter (this is deeper than the left and there is an anatomic barrier created by the phrenicocolic ligament)

RADIOLOGICAL FEATURES

CT Attenuation values range between 0 and +30HU (>30HU with increasing protein content or haemoperitoneum)

- *Loculated peritoneal fluid*: this is due to benign or malignant adhesions ► it appears as a cystic lesion with mass effect

INTRAPERITONEAL AIR

DEFINITION

- This can be caused by a perforation of a hollow viscus, abdominal trauma, surgery or infection

RADIOLOGICAL FEATURES

CT This is able to detect minute quantities of free air ► free air is most commonly seen anterior to the liver (if the patient is supine)

PERITONEAL INFECTION

Peritoneal abscess

- A localized collection of pus within the peritoneal cavity

CT It initially appears as a mass of soft tissue attenuation – it then undergoes liquefactive necrosis with a mature abscess demonstrating wall enhancement and a near water attenuation centre (together with obliteration of the adjacent fat planes) ► gas within a loculated fluid collection is not pathognomonic for an abscess (a necrotic non-infected tumour or mass communicating with the bowel may contain air)

Peritonitis

- A generalized collection of intraperitoneal fluid occurring secondary to bacterial, granulomatous or chemical causes (bacterial peritonitis may be primary or secondary to an intraperitoneal abscess or due to rupture of a hollow viscus)

CT Ascites ► peritoneal (± mesenteric) thickening

Tuberculous peritonitis

- This is rare and can be caused by rupture of a caseous lymph node or direct GI tract involvement by disease, lymphatic or haematogenous spread

CT High attenuation proteinaceous ascites (20–45HU) ► thickening and nodularity of the peritoneal surfaces ► enlarged low attenuation lymph nodes

PERITONEAL FIBROSIS

DEFINITION

Minor peritoneal fibrosis occurs in all patients on continuous ambulatory peritoneal dialysis ► sclerosing peritonitis may develop in a minority

CT Peritoneal thickening ► peritoneal calcification ► loculated fluid collections ► small bowel tethering

INFARCTION OF OMENTUM OR EPIPLOIC APPENDAGE (EPIPLOIC APPENDAGITIS)

DEFINITION

- This occurs either as a result of torsion or from a spontaneous venous thrombosis ► it is a benign, self-limiting condition presenting with acute abdominal pain
  - *Epiploic appendages:* small pouches of peritoneum filled with fat and situated along the colon and upper part of the rectum

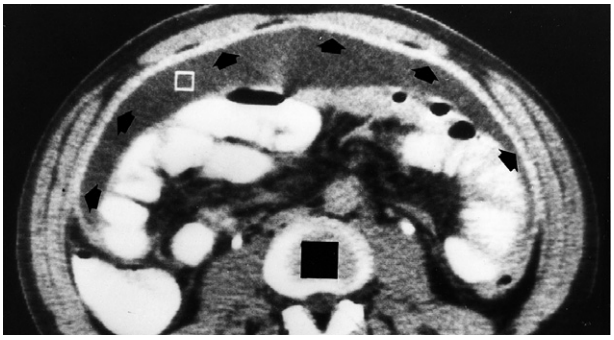
RADIOLOGICAL FEATURES

US An ovoid non-compressible mass of high reflectivity situated under the abdominal wall

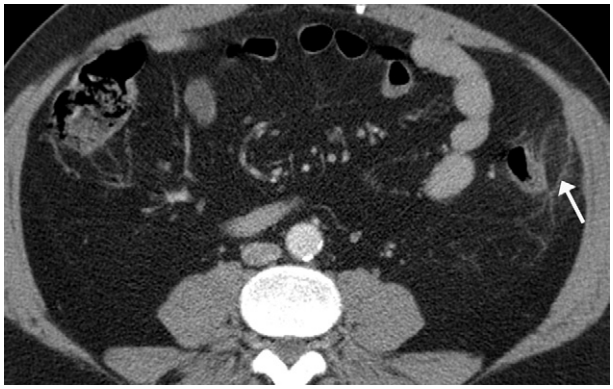
CT A circumscribed fatty area with high attenuation streaks

- In the case of epiploic appendagitis the lesion is seen in contact with the serosal surface of the colon (and usually exhibits a hyperattenuating rim and a central area of high attenuation corresponding to the thrombosed vessels)
- It is also associated with mild local bowel wall thickening

Sclerosing peritonitis. CECT shows a loculated fluid collection and extensive peritoneal calcification.*

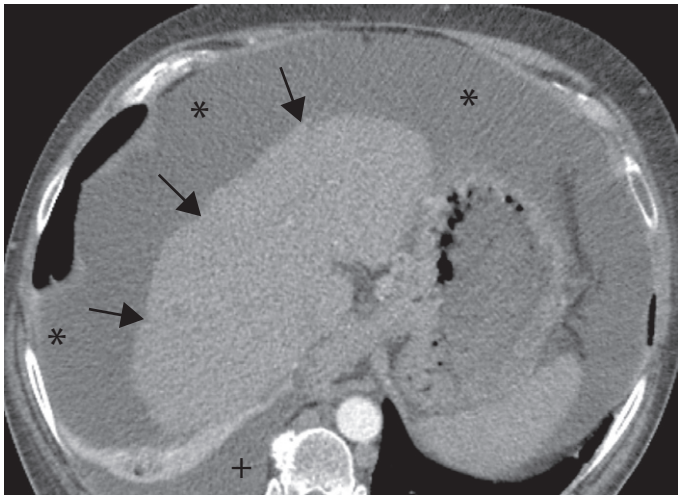


Peritoneal tuberculosis. CT demonstrates marked thickening and enhancement of the parietal peritoneum (arrows). Note high attenuation ascites (cursor).*

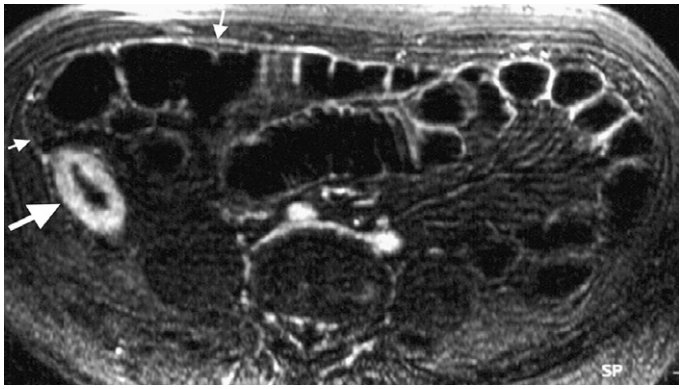


CECT showing an area of increased soft tissue attenuation and stranding of the pericolic fat (adjacent to the descending colon) in keeping with epiploic appendagitis.*

Axial CT demonstrating free intraperitoneal air outside of the bowel lumen.



Ascites. The cirrhotic liver has an irregular edge (arrows) and is surrounded by ascites (*). A right pleural effusion with some collapsed lung is also evident (+).



Peritoneal tuberculosis. T1WI + Gad (FS) depicting enhancement of the peritoneal lining (small arrows). There is involvement of the caecum characterized by homogeneous enhancement of the bowel wall (large arrow).*

NEOPLASTIC PERITONEAL DISORDERS

The majority of peritoneal neoplasms are malignant, and usually ***secondary*** to:

- **Direct invasion**
- **Intraperitoneal seeding (peritoneal carcinomatosis)**
  - **Definition:** malignant tumour seeding of the peritoneum
  - Anywhere where ascites pools will favour malignant growth, therefore the most common seeding sites are: the pouch of Douglas ► the distal small bowel mesentery (near the ileocaecal junction) ► the sigmoid mesocolon ► the greater omentum ► the right paracolic gutter

    CT Sensitivity is reduced for tumour implants < 1cm in diameter
    - Smooth nodular (or plaque-like) thickening and contrast enhancement of the parietal peritoneal surfaces of the diaphragm, liver and spleen (this can also be seen with tuberculosis, peritoneal mesothelioma and peritoneal lymphomatosis)
    - Nodular tumour implants on the undersurface of the right diaphragm can indent the liver surface (mimicking capsular or subcapsular liver metastases)
    - Ascites is not always present – if it is present it is often loculated and septated (and therefore absent from any dependent areas)
  - Calcified peritoneal implants seen pre-chemotherapy suggests that the primary site is usually a serous papillary cystadenocarcinoma of the ovary (or rarely a gastric carcinoma)
  - **Pseudomyxoma peritonei:** this follows rupture of a mucinous cystadenocarcinoma or cystadenoma of the ovary or appendix ► ascites (with septations representing mucinous nodules) and scalloping of the liver edge can be seen
    - **CT:** low attenuation masses
    - **MRI:** T2WI: moderately high SI masses
- **Lymphatic or embolic haematogenous spread**

Primary malignancy	Organ directly invaded	Route of invasion*
Stomach	Spleen	Gastrosplenic ligament
	Superior margin of the transverse colon	Gastrocolic ligament
Pancreas	Liver	Hepatoduodenal ligament
	Inferior margin of the transverse colon	Transverse mesocolon
	Spleen	Splenorenal ligament
Ovary	Diffuse spread through all adjacent peritoneal surfaces	

*Early peritoneal invasion is manifested as linear strands in the fat adjacent to the primary tumour

NEOPLASTIC OMENTAL DISORDERS

Definition

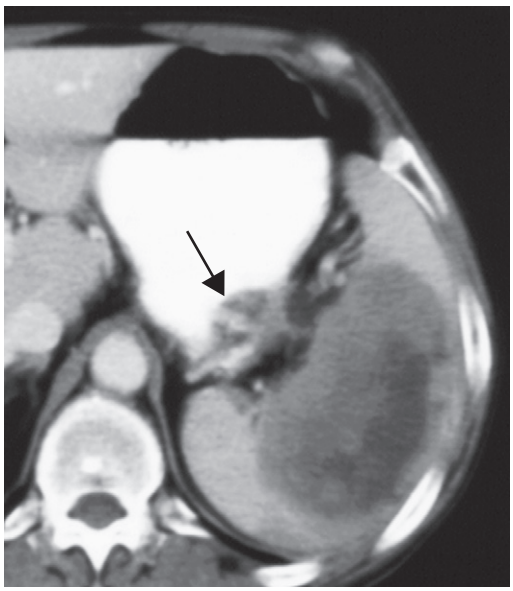
- **Primary neoplasms**
  - These are similar to those encountered within the mesentery
    - Benign neoplasms are usually well circumscribed and localized within the omentum
    - Malignant neoplasms frequently have indistinct margins and infiltrate any surrounding structures
- **Secondary neoplasms**
  - These are more common than a primary neoplasm
    - Tumors metastasizing to the omentum are similar to those responsible for peritoneal carcinomatosis (and usually an ovarian primary)
    - Metastatic disease may involve the greater omentum by direct spread along the transverse mesocolon, gastrosplenic or gastrocolic ligaments (as well as by peritoneal or haematogenous spread)

Radiological features

CT

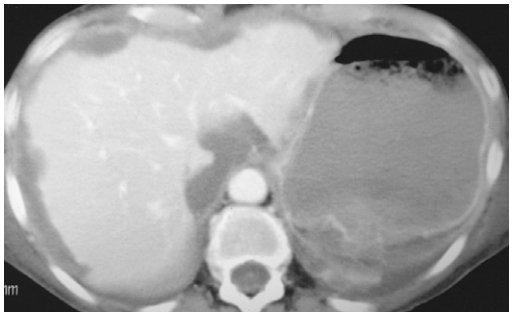
- *Early omental involvement:* irregular soft tissue permeation of the omental fat
- *Advanced omental involvement:* deposits range from discrete nodules to thick, confluent solid omental masses (omental 'cake') ► they may demonstrate enhancement

MRI T1WI: low SI areas within the high SI omental fat ► extensive involvement presents as an intermediate SI crescent-shaped mass ► T1WI + Gad: diffuse enhancement ► images are improved with fat suppression

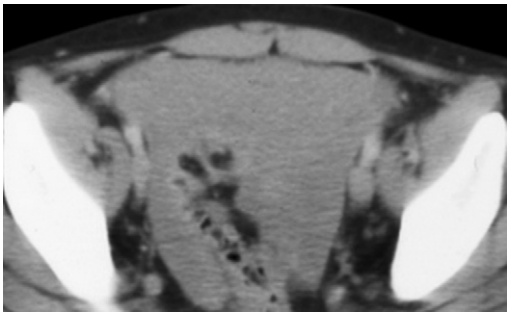


Direct spread of primary splenic lymphoma across the gastrosplenic ligament. The splenic lymphomatous mass has extended along the gastrosplenic ligament to invade the splenic hilar fat and produce mural thickening of the greater curvature of the stomach (arrow).*

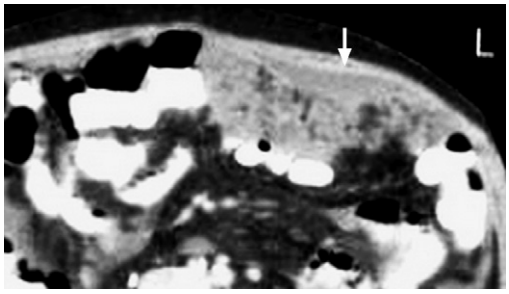
Direct extension of gastric carcinoma across the gastrocolic ligament. A greater curvature carcinoma (white arrow) has spread inferiorly along the gastrocolic ligament to the anterior surface of the transverse colon (black arrows).*



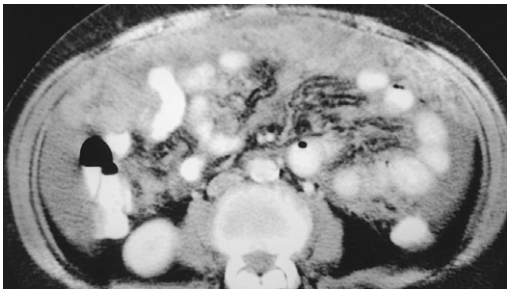
CT reveals the liver scalloping typical of pseudomyxoma peritonei.†



Peritoneal mesothelioma. CECT shows a soft tissue mass that obliterates the pelvic peritoneal spaces and engulfs the sigmoid colon.*



Omental involvement in peritoneal carcinomatosis. CECT of a patient with colonic carcinoma shows confluent soft tissue masses in the distribution of the greater omentum between the anterior abdominal wall and opacified bowel. The 'woven' pattern of masses is caused by entrapped omental fat. There is also loculated ascites (arrow).



CECT reveals plaques of high attenuation peritoneal deposits in a patient with disseminated colorectal adenocarcinoma.

ROTATIONAL ANOMALIES OF THE SMALL BOWEL MESENTERY

DEFINITION

- Rotational anomalies around the axis of the superior mesenteric artery occur when the normal process of fetal gut development is arrested
- It is characterized by the reversal of the normal relationship between the superior mesenteric artery and vein. The artery is now located to the right of vein ► there is twisting of the mesentery around the artery ► there is an absence of a normal horizontal duodenum
- It is usually asymptomatic in adults

DEVELOPMENTAL DEFECTS

DEFINITION

- Internal herniation occurs when the bowel and its mesentery can herniate into pouches or openings within the visceral peritoneum
  - *Paraduodenal hernia*: this is the commonest type and is caused by small bowel entrapment under the right or left mesocolon
  - *Right-sided paraduodenal hernia*: imaging findings include encapsulated small bowel loops within the right mid-abdomen with anterior displacement of the right colic vein, looping of the small intestine around the superior mesenteric vessels, and an abnormal position of the superior mesenteric vein relative to the artery
  - *Left-sided paraduodenal hernia:* the bowel becomes entrapped behind the descending mesocolon within the paraduodenal fossa with anterior displacement of the inferior mesenteric vein by the dilated encapsulated bowel loop

LYMPHANGIOMA

DEFINITION

- The commonest subtype of a mesenteric cyst – it represents a congenital malformation of the bowel lymphatic vessels, frequently surrounding the loop of bowel from where it originates

RADIOLOGICAL FEATURES

US This can demonstrate internal septations

CT A large, thin-walled, single or multiloculated cystic mass ► its contents are of water-to-fat attenuation

MRI T2WI: high SI

ENTERIC DUPLICATION CYST

DEFINITION

- An uncommon congenital anomaly found anywhere along the GI tract (commonly within the ileum) and located on the mesenteric border
- It is lined with alimentary tract mucosa (occasionally gastric or pancreatic mucosa)

RADIOLOGICAL FEATURES

US Its wall is thick and composed of multiple layers, like those of the normal bowel wall

CT/MR A unilocular mass of predominantly water content and a thick wall that exhibits contrast enhancement

MESOTHELIAL CYST

DEFINITION

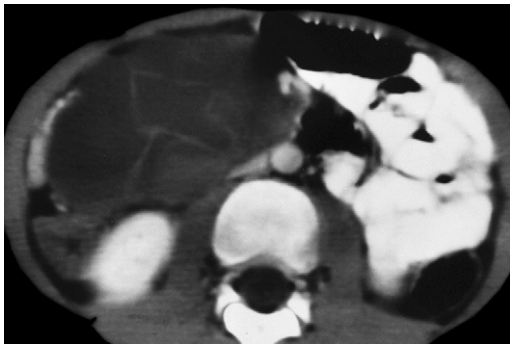
- This results from failure of coalescence of mesothelial-lined peritoneal surfaces

RADIOLOGICAL FEATURES

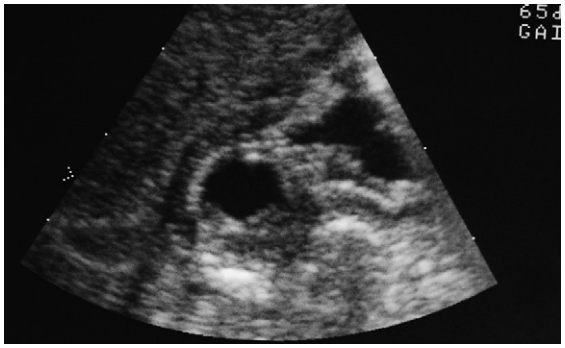
CT A fluid-filled mass with no discernible wall ► no internal septations are demonstrated (cf. a lymphangioma)

Small bowel loops are in the right upper quadrant in this patient with malrotation.••

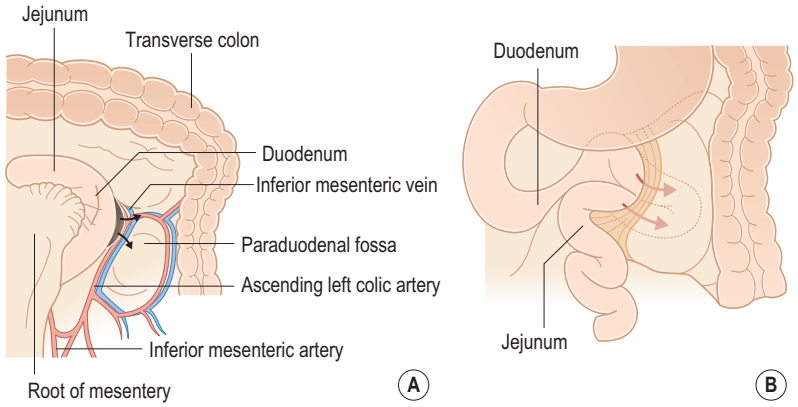
Typical corkscrew appearance of the duodenum and proximal jejunum associated with malrotation and midgut volvulus.©20



Cystic mesenteric lymphangioma. Enhanced CT depicting a multilocular cystic mass with thin internal septa, occupying the small bowel mesentery.*



Duplication cyst of pylorus. Ultrasound showing echogenic mucosal layer and hypoechoic outer muscular layer.*



Development of a left-sided paraduodenal hernia. (A) Small bowel loops herniate into the descending mesocolon through the paraduodenal fossa posterior to the inferior mesenteric vein and ascending left colic artery. (B) Small bowel loops progressively herniate through an abnormal peritoneal pocket.

Mesenteric lymphadenitis

Definition
- Benign lymph node inflammation within the ileal mesentery ► clinically it can mimic an appendicitis

US/CT Moderately enlarged mesenteric lymph nodes within the right lower quadrant (± ileal or ileocaecal wall thickening)

Small bowel perforation

Definition Perforation that is not associated with blunt trauma (e.g. a ruptured diverticulum or an intramural foreign body penetrating the wall)

CT Streaky mesenteric soft tissue densities associated with local extraluminal gas

Graft-vs-host disease

Definition A complication of heterotopic bone marrow transplantation

CT Focal or diffuse mural bowel thickening (usually within the ileum) ► it is associated with an increased size of any associated mesenteric vessels

Whipple's disease

Definition A systemic infectious disease primarily associated with malabsorption

CT Low attenuation mesenteric and retroperitoneal lymph nodes (due to the nodal deposition of fat and fatty acids) ► it is associated with diffuse intestinal wall thickening

Mesenteric panniculitis

Definition Chronic non-specific inflammation involving the small bowel mesenteric adipose tissue ► a rare, slowly progressive condition of unknown origin
- *Retractile or fibrosing mesenteritis:* a dominant fibrotic component is present

CT (mesenteric panniculitis) A well-delineated, inhomogeneous fatty mass located at the mesenteric root ► there is an absence of adjacent bowel loop involvement (or bowel displacement) ► there is mesenteric vascular envelopment (with a low attenuation halo surrounding the vessels)

CT (retractile mesenteritis) An infiltrative soft tissue mass with associated radiating linear strands of soft tissue attenuation (which may mimic a desmoid or carcinoid tumour) ► calcification may be present within the necrotic central portion of the mass

MRI Fibrosing mesenteritis: T1WI/T2WI: low SI

Non-inflammatory oedema

Definition A diffuse increase in mesenteric attenuation obscuring the mesenteric vessels
- *Causes:* hypoalbuminaemia ► cirrhosis ► nephrotic syndrome ► right-sided congestive heart failure ► mesenteric ischaemia ► vasculitis ► trauma

CT (mesenteric ischaemia) Mesenteric oedema can be focal or diffuse depending on the extent of vascular compromise
- Circumferential radially symmetric (<1.5cm) bowel wall thickening ► increased mesenteric fat attenuation (secondary to oedema) ► decreased, delayed, or lack of small bowel enhancement ► intramural, mesenteric or portal venous gas with bowel infarction ► a low-density thrombus may be seen within the proximal mesenteric arteries

Radiation

Definition Radiation produces an endarteritis within the radiation port

CT Linear streaks of increased attenuation within the mesenteric fat (representing oedema) ► mesenteric retraction ► progressive small-vessel narrowing and vascular congestion ► bowel wall fibrosis ► thick-walled and straightened bowel loops

Mesenteric lymphoedema

Causes
- *Lymphatic obstruction:* this is secondary to inflammation, surgery, or a neoplasm
- *Congenital malformations of the lymphatic system:* collateral flow can occur via the mesenteries with secondary lymphoedema
- *Intestinal lymphangiectasia:* lymphatic stasis with mesenteric lymphoedema and chylous ascites
- *Metastatic disease:* lymph node involvement at the root of the small bowel mesentery may cause central lymphatic obstruction

Mesenteric and small bowel injuries

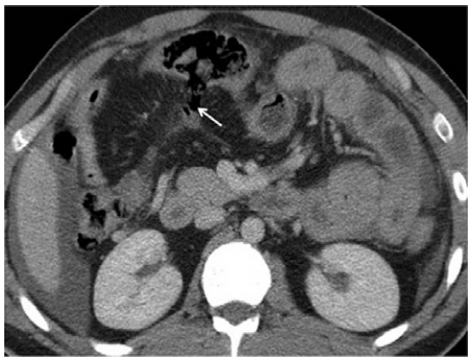
Definition An uncommon sequelae of blunt abdominal trauma

CT Free air and contrast extravasation (a highly specific sign – however their absence does not exclude a bowel wall injury) ► intestinal wall thickening (a non-specific sign) ► streaky densities within the mesenteric fat ► a mesenteric haematoma ► a triangle-shaped fluid collection within the mesentery
- *'Sentinel clot' sign:* the portion of blood closest to the source of bleeding may demonstrate the highest attenuation values (due to clot formation close to the bleeding source)

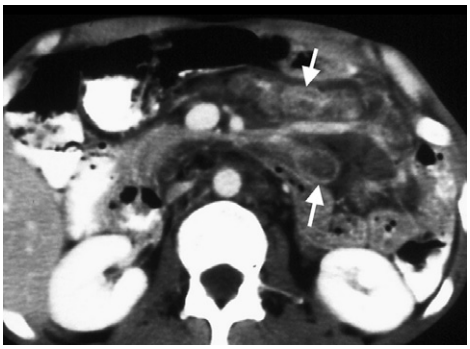
Intestinal obstruction

CT (strangulating obstruction) Engorgement of the mesenteric vessels ► haziness of the mesenteric fat ► blurring of the margins of the vessels and mesenteric fluid

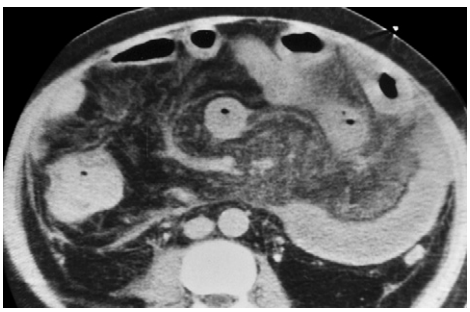
CT (closed-loop obstruction) Any mesenteric involvement is less dramatic, with convergence of the dilated veins toward the point of obstruction



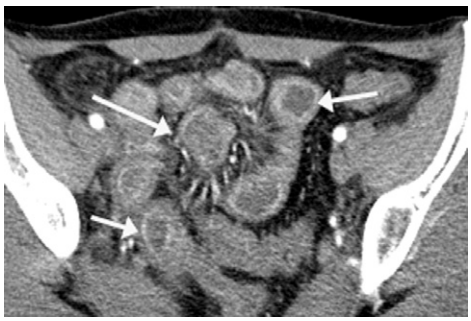
Small bowel perforation. CECT shows abnormally thickened loops of small bowel with adjacent free gas (arrow) in a patient with Crohn's disease.*



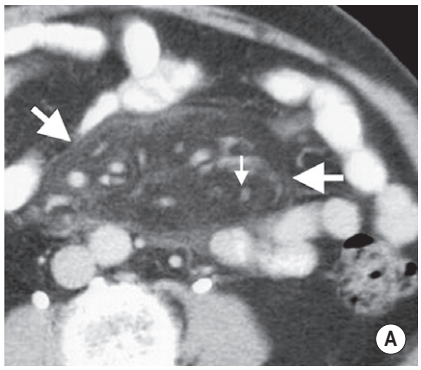
Whipple's disease. Non-enhancing low attenuation lymph nodes are seen within the small bowel mesentery (arrows).

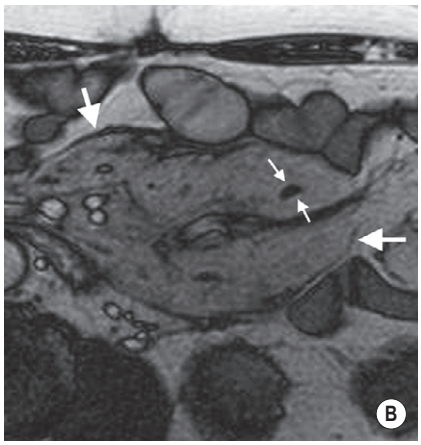


Mesenteric ischaemia. NECT shows diffuse haziness of the affected small bowel mesentery with effacement of the vascular markings, reflecting oedema, haemorrhage and venous congestion. Note also the thickened high attenuation bowel wall due to intramural haemorrhage.*

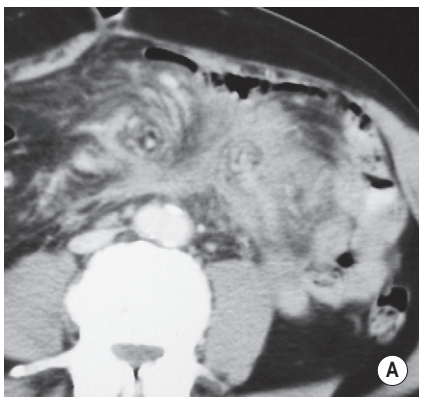


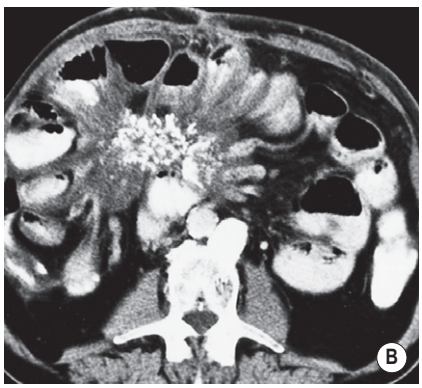
CECT showing multiple mildly thickened abnormally enhancing loops of small bowel (arrows) in a patient with previous pelvic radiotherapy for cervical cancer.*





Mesenteric panniculitis. CECT (A) and true-FISP MRI (B) in a patient who presented with abdominal pain show a well-delineated fatty mass (large arrows) extending from the root of the small bowel mesentery toward the left abdomen, engulfing mesenteric vessels without distortion. Note the perivascular halo (small arrow).*





Fibrosing mesenteritis. (A) CECT in a patient who presented with fever of unknown origin demonstrates a fibrofatty mesenteric mass with irregular borders surrounding mesenteric vessels. Strands of soft tissue density are seen radiating from the mass to the adjacent mesenteric fat. (B) CT appearances. CECT demonstrating a large, ill-defined, soft tissue mesenteric mass with extensive calcification. Note retraction and thickening of the adjacent bowel loops.

NEOPLASTIC MESENTERIC DISORDERS

Definition

- Primary neoplasms arising within the mesentery are rare and are usually of mesenchymal origin
  - Benign primary mesenteric tumours (e.g. fibromatosis, lipoma, neurofibroma) are more common than malignant tumours (e.g. fibrosarcoma, liposarcoma, mesothelioma)
  - Secondary neoplasms are more common than primary malignancies
  - Cystic tumours are more common than solid tumours
  - Malignant tumours tend to be located near the mesenteric root ► benign tumours tend to arise within the periphery (near the bowel wall)
- *The most common malignancies spreading into the mesentery:* metastatic carcinoma ► lymphoma ► carcinoid tumour

PRIMARY NEOPLASMS

Malignant peritoneal mesothelioma

- The commonest primary peritoneal neoplasm (occurring in middle-aged men) ► it is associated with asbestos exposure (there is not always pleural involvement) ► it has a poor prognosis

CT Diffuse thickening or nodularity of the peritoneum ► it may diffusely infiltrate the mesentery, leading to omental and mesenteric thickening (with a 'stellate' configuration of the thickened perivascular bundles or 'pleated' thickening of the mesenteric leaves) ► peritoneal and omental masses (± local invasion into adjacent organs)

- Any ascites is disproportionately small (cf. large-volume ascites with metastatic disease)

Cystic mesothelioma

A rare benign neoplasm that is not associated with asbestos exposure ► it is frequently seen within the pelvis and appears as a unilocular or complex cystic mass (stimulating a lymphangioma or ovarian carcinoma)

Fibromatosis

- The commonest primary solid mesenteric tumour ► it can occur in an isolated form or can be associated with Gardner's syndrome ► it has a tendency to arise following either surgery or trauma

CT One or more usually large soft tissue attenuation masses (>10cm) ► it often has irregular or ill-defined margins ► tethering, encasement or invasion of the adjacent bowel can be seen ► it usually demonstrates greater enhancement than muscle

Lipoma

- The 2nd commonest primary solid mesenteric tumour

CT A well-circumscribed homogeneous mass of fat attenuation

MRI T1WI/T2WI: high SI ► T1WI (FS): low SI ► internal septations are unusual

Liposarcoma

- This is more commonly found within the retroperitoneum than within either the mesentery or peritoneum

CT/MRI A variable appearance reflecting its tissue composition – it can range from predominantly fat, fluid and soft tissue elements to an entirely soft tissue density mass

SECONDARY NEOPLASMS

Metastatic carcinoma

- The primary is usually the stomach, colon or ovary

Intraperitoneal tumour dissemination The small bowel mesentery is frequently involved by intraperitoneally disseminated tumour with non-specific findings:

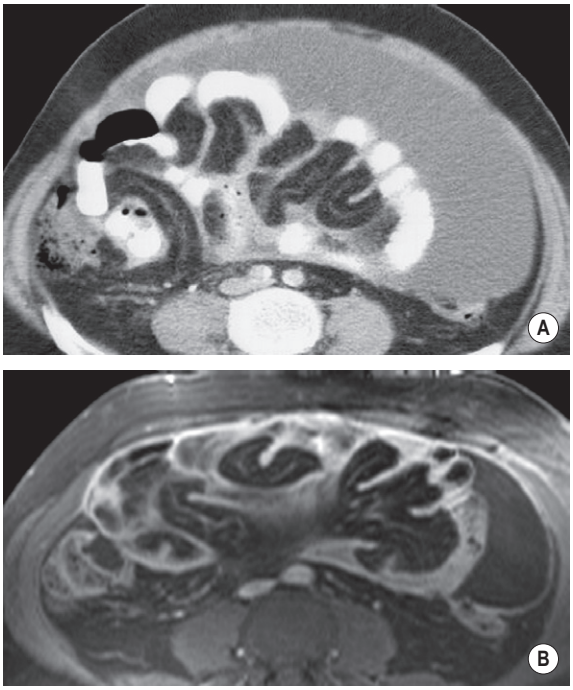
- Scattered nodules ► rounded, ill-defined soft tissue or cystic masses
- Any ascites can be loculated, and if large enough will tend to surround bowel loops which are tethered centrally by the rigid mesentery
- Diffuse mesenteric infiltration may resemble mesenteric oedema
- Metastatic tumour nodules on the visceral peritoneal surfaces can become adherent to the serosa of the small bowel loops
- A severe desmoplastic response to the seeded metastases can cause marked fixation and angulation of the ileal loops (± obstruction)
- Fixation and thickening of the mesentery:
  - *Stellate form:* a radiating configuration of the mesenteric folds with thickened rigid perivascular bundles and encased, straightened vascular structures
  - *Pleated appearance:* sheets of soft tissue produce thickening of the mesenteric folds

Embolic metastases These spread via the mesenteric arteries to locate along the antimesenteric border of the small bowel (e.g. melanoma, lung, or breast primaries)

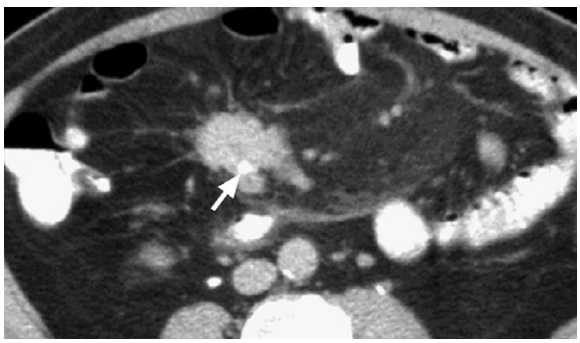
- **CT:** focal bowel wall thickening ► thickening of the mesenteric folds ► melanoma deposits may become large and ulcerated ► breast cancer deposits may cause multiple areas of small bowel luminal narrowing with prestenotic dilatations

Lymphatic dissemination This plays a minor role in the spread of metastatic carcinoma but it is the main pathway of dissemination of lymphoma to the mesenteric lymph nodes ► enlarged mesenteric lymph nodes occur at presentation in approximately 50% of patients with NHL

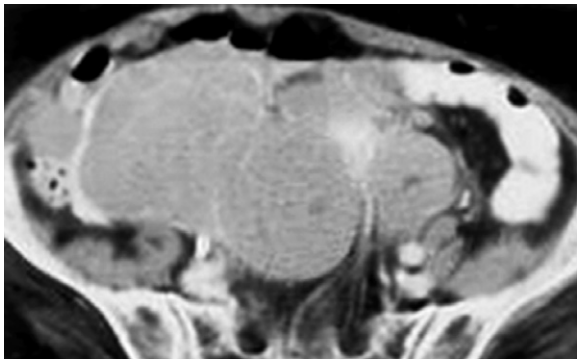
- **CT:** confluent lymphomatous nodes may surround the superior mesenteric vessels producing a 'sandwich-like' appearance ► coexisting lymphomatous mural involvement of the small bowel loops will affect their mesenteric border



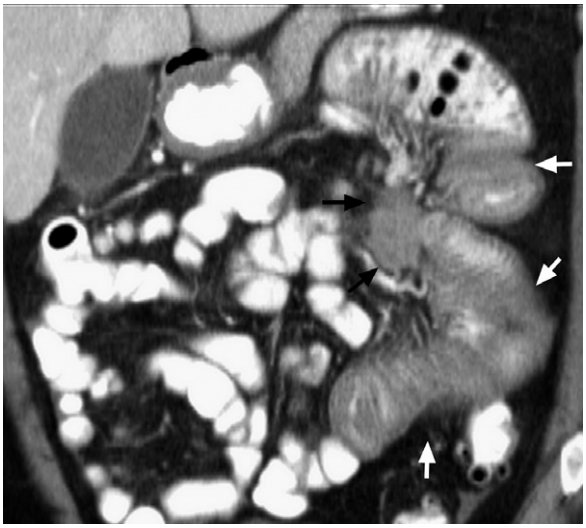
Seeded gastric carcinoma along the small bowel mesentery. CECT (A) and post-gadolinium T1-weighted MR image (B) show thickening and enhancement of the mesentery together with ascites.*



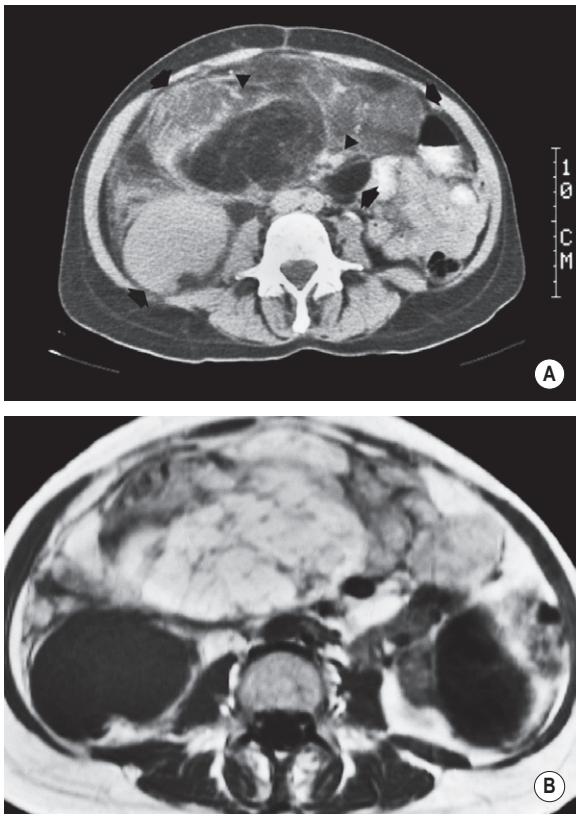
Carcinoid. CECT shows a mesenteric mass with radiating strands toward adjacent bowel loops. An area of dystrophic calcification is also evident (arrow).*



Mesenteric fibrosarcoma. CECT shows an enhancing soft tissue mass in the small bowel mesentery compressing a neighbouring small intestinal loop.*



Mesenteric fibromatosis. CECT shows a soft tissue mass in the mesentery (black arrows) resulting in segmental jejunal ischaemia, manifested as symmetrical wall thickening (white arrows).*



Liposarcoma: CT-MR pathological correlation. (A) CECT of the abdomen demonstrates a large mesenteric mass (arrows) with both fat and soft tissue densities, encasing large peripheral vessels (arrowheads). (B) T1WI demonstrating the multiple fibrous strands seen in this liposarcoma, as well as two components, one brighter centrally (corresponding to a fatty element) and a lower-intensity peripheral component (corresponding to an undifferentiated sarcomatous element).*

3.11 PLAIN ABDOMINAL RADIOGRAPH

PNEUMOPERITONEUM

DEFINITION

- Free intra-abdominal gas – it usually indicates perforation of a viscus (it is often a peptic ulcer – a perforated appendix rarely demonstrates free gas)
  - *Other causes:* bowel obstruction ► appendicitis ► bowel ischaemia ► diverticular disease ► post colonoscopy

RADIOLOGICAL FEATURES

Erect CXR Gas is seen under the diaphragm (this can detect as little as 1ml of free gas) ► do not confuse this appearance with Chilaiditi's syndrome (where intestine is seen between the liver and diaphragm) or a subphrenic abscess

Left lateral decubitus AXR Gas is seen between the liver and abdominal wall

Supine AXR Gas is seen within the RUQ – particularly within the subhepatic space and hepatorenal fossa (Morrison's pouch) ► triangular collections of air are seen within the abdomen (outlining the visceral contents) ► gas is seen on either side of the falciform ligament ► scrotal air can be seen in children

- *'Inverted V' sign*: gas is seen on either side of the umbilical ligaments
- *'Rigler's sign:* the outer and inner walls of a bowel loop are delineated by gas
- *'Football' or 'air dome' sign:* a central round air collection seen on a supine AXR in children (as air rises)

CT (lung window settings) This is the most sensitive technique for detecting small amounts of free gas (look anterior to the liver, anteriorly within the central abdomen, and within the peritoneal recesses)

GAS WITHIN THE RETROPERITONEUM

DEFINITION

- Retroperitoneal gas can be seen particularly if the originating organ is retroperitoneal
  - *Causes:* a perforated posterior peptic ulcer ► a perforated sigmoid diverticular disease ► post colonoscopy

RADIOLOGICAL FEATURES

AXR/CT Gas is seen within the layers of the abdominal wall (flanks) or around the kidneys ► gas can track superiorly into the mediastinum and inferiorly into the buttock and thigh (classically gas is seen within the soft tissues of the left thigh from a diverticular perforation)

GAS WITHIN THE BOWEL WALL

Causes Intestinal infarction (following thrombosis or embolism of the superior mesenteric artery) ► pneumatosis cystoides intestinalis

Intestinal infarction

AXR/CT Linear gas streaks seen within the bowel wall ► non-specific dilated loops of small bowel ► thickening of the small bowel wall (due to submucosal haemorrhage or oedema) ► free gas (if there has been a perforation) ► mesenteric or portal venous gas (in advanced cases)

Pneumatosis cystoides intestinalis (pneumatosis coli)

Definition Cyst-like collections of gas within the submucosal or subserosal layers of the bowel wall (there is a normal overlying mucosa) ► the cysts can occasionally rupture, producing a pneumoperitoneum ► causes:

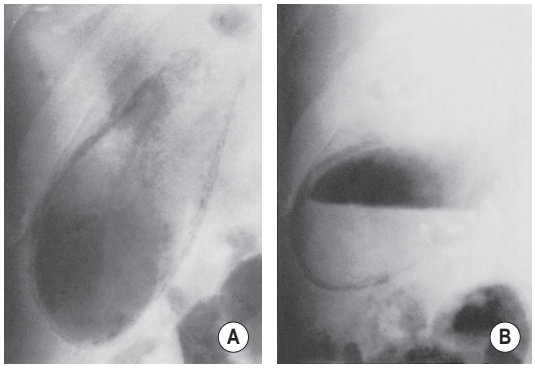
- *Pulmonary disease:* air tracks along the lung interstitium, via the mediastinum, to the retroperitoneum and mesentery (there is a known association with COPD)
- *Bowel necrosis:* this is seen with necrotizing enterocolitis and mesenteric thrombosis
- *Mucosal disruption:* this is seen with intestinal obstruction or trauma (e.g. endoscopy)

AXR/CT The cysts are well defined and closely packed (1–3cm in diameter) ► they usually affect the left hemicolon

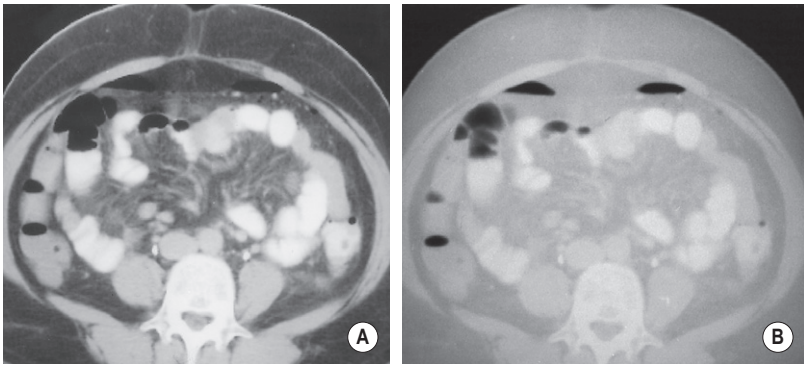
Treatment Prolonged high-dose oxygen therapy (the resultant altered diffusion gradients will collapse any cysts)

Gas within the wall of other organs	
Within the biliary tree	Following a gallstone ileus
Gallbladder	With emphysematous cholecystitis
Portal veins	Secondary to mesenteric infarction
Kidneys	Following emphysematous pyelonephritis
Pancreas	With infected necrosis or abscess
Urinary bladder	With infection or following catheter insertion

Causes of pneumoperitoneum without peritonitis*
Silent perforation of viscus that has sealed itself, in:
Patients on steroids
Unconscious patients
Patients being ventilated
The presence of other serious medical conditions
Postoperative
Peritoneal dialysis
Perforated cyst in pneumatosis cystoides intestinalis
Tracking down a pneumomediastinum
Stercoral ulceration
Leakage through a distended stomach (e.g. endoscopy)
Vaginal-tubal entry of air

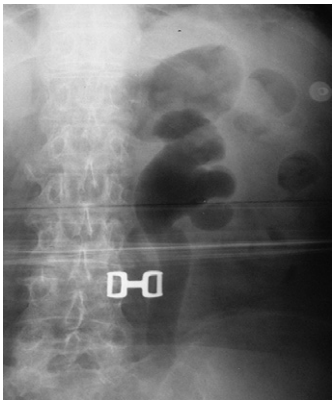


Emphysematous cholecystitis showing (A) gas in the lumen and wall of the gallbladder and (B) a gas–fluid level in the erect posture.†

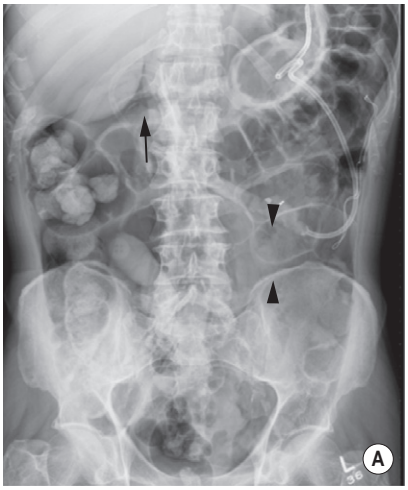


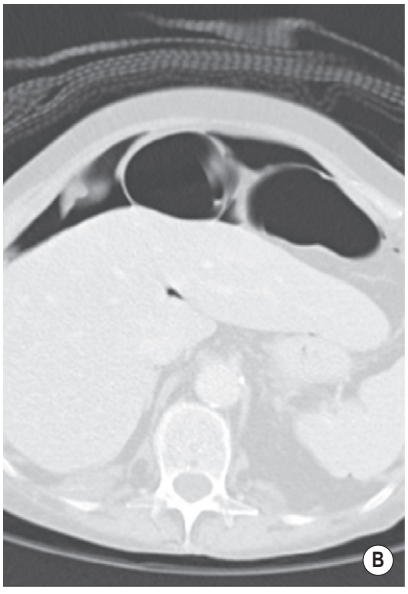
Free peritoneal gas due to perforated sigmoid diverticular disease. (A) CT viewed on abdominal window settings. (B) The same image viewed on broad window settings. The free gas deep to the anterior abdominal wall is more conspicuous in (B).*

Pneumatosis coli. Multiple small gas-filled cysts are seen in association with the colon. There is also free peritoneal gas.*



Emphysematous pyelonephritis. Patient with diabetes mellitus and sepsis. The left renal collecting system and ureter are distended and gas filled. There are also multiple dense gallstones in the gallbladder.*





Rigler's sign. (A) Both sides of bowel wall are outlined due to gas being present internally and externally (arrowheads). In addition, triangular collections of air (arrow) are seen. (B) CT (lung windows) confirms a large amount of free intraperitoneal air.

Pneumatosis coli with numerous gas cysts in the wall of the colon.*

Retroperitoneal gas due to perforated sigmoid diverticular disease. Film from an IVU series. There is gas between the layers of the abdominal wall and around the upper pole of the left kidney.*

OMPHALOCELE (EXOMPHALOS)

DEFINITION

- Incomplete formation of the embryonic ventral abdominal wall leads to a congenital midline anterior abdominal wall defect around the umbilicus (the umbilical cord inserts at the tip of the defect)
  - *Larger omphaloceles (containing liver tissue):* due to failure of lateral body fold fusion
  - *Smaller omphaloceles (containing bowel only):* due to persistence of physiological gut herniation
- Associated chromosomal abnormalities are common (50%): trisomy 13 and 18
  - *Beckwith–Wiedemann syndrome*: omphalocele (exomphalos) + macroglossia + gigantism (the 'EMG' syndrome) ► visceral abnormalities are seen in up to 70% of cases
- **Diagnosis:** antenatal US

GASTROSCHISIS

DEFINITION

- A small defect in the ventral abdominal wall, classically to the right side of a normally positioned umbilicus ► due to a localized intrauterine vascular accident leading to focal full-thickness necrosis of the anterior abdominal wall
  - Typically no other anomalies

RADIOLOGICAL FEATURES

Antenatal US Bowel loops floating freely in the amniotic fluid with no covering membrane

- Exposure to the amniotic fluid damages the bowel and the extruded bowel loops are dilated and thickened

Postnatal complications Necrotizing enterocolitis (NEC) in up to 20% ► respiratory embarrassment following repair of the defect ► short bowel syndrome and intestinal dysmotility ► intestinal atresias and stenoses (secondary to the prenatal ischaemic insult)

- The associated morbidity and mortality is mainly due to the associated GI problems
- Approximately ⅓ of males have cryptorchidism, which may result in the testes passing through the defect

CLOACAL EXSTROPHY

DEFINITION

- A rare midline infra-umbilical defect that arises due to an abnormality of the caudal body fold (M>F):
  - Omphalocele + an imperforate anus + spinal dysraphism + ambiguous genitalia ► the caecum opens onto the anterior abdominal wall between the two exstrophied hemi-bladders

RADIOLOGICAL FEATURES

AXR The pubic bones are separated by > 25mm

Upper GI contrast study This can detect the presence of malrotation and also outlines the length of bowel present

MRI This can exclude an associated cord anomaly and delineate the pelvic organs and the pelvic floor musculature

TREATMENT

- The bladder and bowel are separated and repaired, with the abdominal wall defect closed
  - Previously, genetically male infants with cloacal exstrophy have commonly undergone a bilateral orchidectomy and been reared as girls (as the external genitalia are frequently rudimentary)
  - Genetically female infants usually have a double vagina and two hemi-uteri

PRUNE BELLY (EAGLE–BARRETT) SYNDROME

DEFINITION

- A non-hereditary disorder consisting of:
  - *A congenital absence of the abdominal wall muscles*: giving a wrinkled and lax abdominal wall
  - *Urinary tract abnormalities*: renal dysplasia + gross pelvicalyceal and ureteric dilatation
  - *Cryptorchidism:* the bladder distension interferes with testicular descent

RADIOLOGICAL FINDINGS

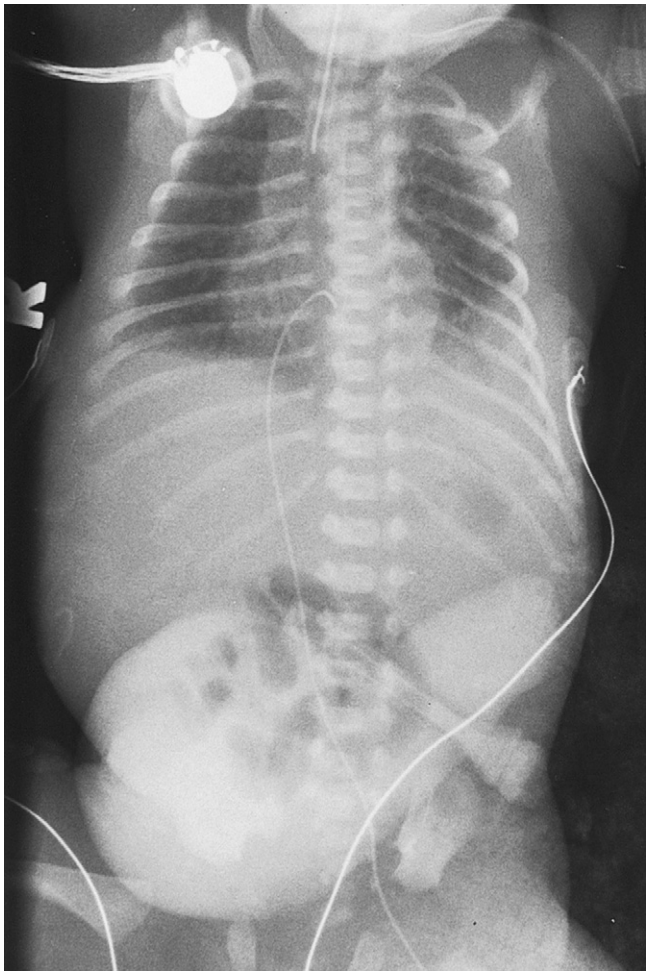
AXR A protuberant abdomen (resulting from the lack of abdominal musculature)

US/micturating cystourethrogram There are small kidneys with abnormal minimally dilated calyces and upper ureters – the lower ureters are tortuous and show disproportionate dilatation ► the bladder is thin walled, of a large capacity (without trabeculation), and has a wide neck ► there may be a patent urachus or a urachal diverticulum ► the posterior urethra is dilated proximally with typical conical narrowing (± dilatation of the anterior urethra)

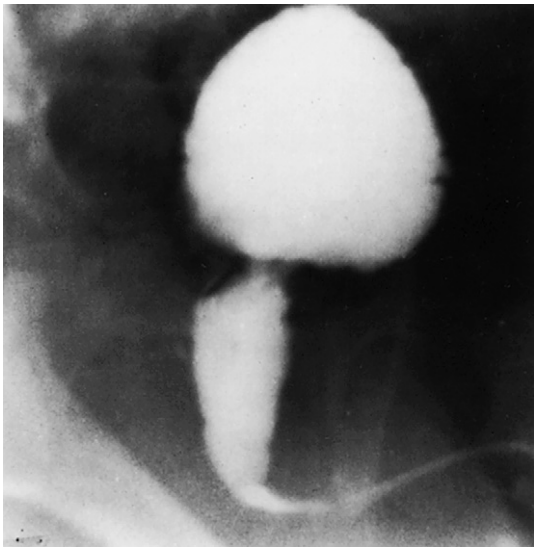
- **MCUG:** this is valuable for assessing the urethral anatomy but is dangerous if sepsis arises ► vesicoureteral reflux is seen in ⅔ of patients

^{99m}Tc-DMSA This assesses the divided renal function

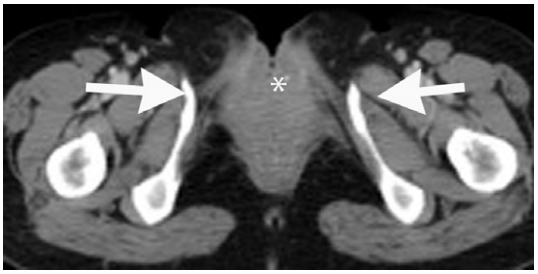
Dynamic renal scintigraphy This fails to show adequate drainage due to the gross dilatation



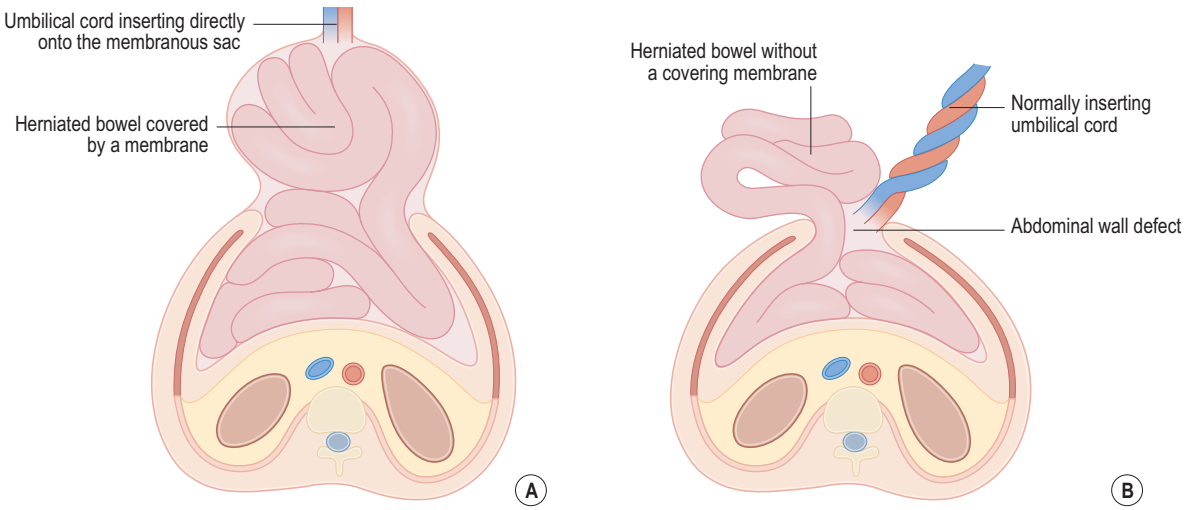
Gastroschisis. Several air-filled extra-abdominal loops of bowel are seen in this infant of 26 weeks' gestation. A small left congenital diaphragmatic hernia was also present.†



Prune belly syndrome. Image exposed near the end of micturition on a voiding cystourethrogram. The posterior urethra is dilated proximal to the membranous urethra and the calibre of the latter is normal. Posterior urethral valves are not present.*



Cloacal exstrophy: axial CECT showing diastasis of the symphysis pubis (arrows) and midline defect in the lower anterior abdominal wall (*).



(A) Omphalocele. A midline abdominal wall defect with herniated bowel covered by a membrane. The umbilical cord inserts directly onto the herniated membranous sac. (B) Gastroschisis. The abdominal wall defect is adjacent to a normally inserted umbilical cord. The herniated bowel is not covered by a membrane.

NON-BILIOUS VOMITING

Definition

- Neonatal non-bilious vomiting due to a GI cause implies the presence of a lesion proximal to the ampulla of Vater and is most frequently due to GOR
  - Congenital gastric obstruction is rare and is usually due to a web or diaphragm within the antrum and pylorus ► occasionally a true atresia is present with a fibrous cord uniting the two blind ends

Radiological findings

Prenatal

US Maternal polyhydramnios ► a large fetal gastric bubble

Postnatal

AXR With complete obstruction there will be a dilated stomach with no distal air

Upper GI series Vigorous gastric peristalsis and consistent filling defects within the antrum or pylorus at the site of the web

- *'Pseudo-double bubble' sign:* this is seen as barium outlines first the space between the antrum and pylorus, and then the duodenal bulb

US Persistent linear, echogenic structures arising from the antral or pyloric walls and extending centrally

BILIOUS VOMITING

Definition

- An obstruction distal to the ampulla of Vater – malrotation and a midgut volvulus constitute the greatest emergency
  - *Other causes:* duodenal atresia and stenosis ► duodenal webs and diaphragms ► extrinsic duodenal compression (e.g. an annular pancreas or a preduodenal portal vein) ► small bowel atresia ► small bowel stenosis
  - If the AXR demonstrates a complete high intestinal obstruction then no further imaging is required ► if the AXR shows a low intestinal obstruction (i.e. distal to the mid ileum) then a contrast enema is preferred

DUODENAL ATRESIA AND STENOSIS

Definition

- This is caused by failure of recanalization of the duodenal lumen after the 6th week of fetal life (duodenal atresia is much more common than a duodenal stenosis) ► in 80% of cases the level of obstruction is just distal to the ampulla of Vater
  - *Associated anomalies occur in the majority of patients:* Down's syndrome (30%) ► malrotation (20–30%) ► congenital heart disease (20%) ► components of the VACTERL association may also be present

Clinical presentation

- Infants present early with bilious vomiting and upper abdominal distension (a preampullary obstruction presents with non-bilious vomiting)

Radiological features

Antenatal

US A dilated stomach and duodenal cap ► maternal polyhydramnios

Postnatal

AXR A gas-filled 'double bubble' of the stomach and duodenal cap ► distal gas will be present if the obstruction is partial (or rarely if there is a bifid pancreatic duct straddling the atretic segment)

Upper GI study A duodenal stenosis is seen as a narrowed area within the 2nd part of the duodenum ► a duodenal web may be seen as a thin, filling defect extending across the duodenal lumen

- *'Duodenal dimple' sign:* the pressure exerted by a NGT on the obstructing web can cause in-drawing of the duodenal wall at the site of the web's attachment

SMALL BOWEL ATRESIA AND STENOSIS

Definition

- These follow an intrauterine vascular insult (the vascular insult may be a primary or secondary event such as an antenatal volvulus or intussusception) ► an atresia is more common than a stenosis ► the proximal jejunum and distal ileum are the most frequently affected segments

Clinical presentation

- The majority of infants present with bilious vomiting in the immediate postnatal period ► abdominal distension is seen with more distal atresias

Radiological findings

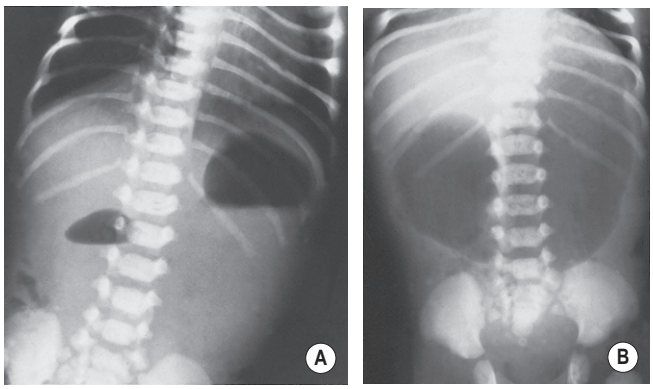
AXR Dilated loops of small bowel are seen down to the level of the atresia (the loop of bowel immediately proximal to the atresia may be disproportionately dilated and have a bulbous contour) ► bubbles of distal gas are seen with a stenosis (cf. an atresia) ► fine intraluminal calcifications may be seen with a more distal atresia ► a meconium peritonitis (with calcification of the peritoneum) may be seen if an intrauterine perforation has occurred

- *'Apple peel' syndrome:* this is thought to follow an intrauterine occlusion of the distal SMA ► there is a proximal jejunal atresia, with agenesis of the mesentery and absence of the mid small bowel ► the distal ileum spirals around its narrow vascular pedicle (giving the syndrome its name) ► a malrotated microcolon is also usually present

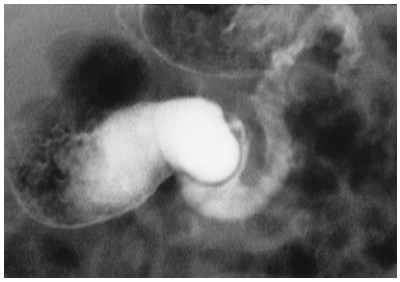
Pearl

- Medical causes of bilious vomiting include functional immaturity of the colon and gastroenteritis

Causes of gastrointestinal obstruction†
Oesophagus
Oesophageal artesia ± tracheal-oesphageal fistula
Congenital oesophageal stenosis, web and diverticula
Extrinsic compression - vascular ring
- foregut duplication cyst
- neoplasm
Stomach
Gastric atresia
Antral web
Duplication cyst
Hypertrophic pyloric stenosis
Duodenum
Duodenal artesia
Duodenal web
Malrotation with midgut volvulus
Extrinsic compression - annular pancreas
- preduodenal portal vein
Small bowel
Jejunal and ileal atresia/stenosis
Meconium ileus ± meconium cyst, segmental volvulus
Midgut volvulus
Inguinal hernia
Necrotizing enterocolitis
Duplication cyst
Large bowel
Hirschsprung's disease
Functional immaturity/hypoplastic left colon syndrome
Colonic atresia/imperforate anus
Necrotizing enterocolitis
Duplication cyst



Duodenal atresia. Erect (A) and supine (B) AXR demonstrating the classic 'double bubble' sign.*



Duodenal web. Barium meal demonstrating a curvilinear filling defect or 'wind-sock diverticulum' in the second part of the duodenum with proximal dilatation.†

Ileal atresia. Contrast enema with microcolon and reflux into a non-dilated distal ileal segment with abrupt convex termination. A few meconium plugs are present.†

Malrotation and volvulus. Upper GI contrast medium study demonstrates the classical 'corkscrew' pattern of the duodenum and jejunum spiralling around the mesenteric vessels.*

MALROTATION

DEFINITION

- Malrotation is a generic term used to describe any variation in the intestinal position ► intestinal malfixation invariably accompanies malrotation in an attempt to fix the gut in place
  - *Peritoneal (Ladd) bands:* these stretch from the abnormally high-lying caecum, across the duodenum, and to the region of the porta hepatis and the anterior and posterior abdominal walls ► Ladd bands can cause duodenal obstruction
- The abnormal positions of the duodenojejunal junction and caecum means that the base of the small bowel mesentery is short
  - *Midgut volvulus:* the midgut has a propensity to twist around this narrow base compromising its vascular supply – this can lead to ischaemic necrosis of the small bowel with an associated high mortality rate if undiagnosed

CLINICAL PRESENTATION

- This commonly presents within the 1st month of life with bilious vomiting ► older children may present with non-specific symptoms of chronic or intermittent abdominal pain, non-bilious emesis, diarrhoea, or a failure to thrive
- Symptoms of shock intervene if bowel ischaemia and necrosis have developed

RADIOLOGICAL FEATURES

AXR There are no specific features – it may be normal if a volvulus is intermittent or if there is incomplete duodenal obstruction due to loose twisting of the bowel

- A tight volvulus results in complete duodenal obstruction with a distended stomach and proximal duodenum (mimicking the 'double bubble' of duodenal atresia)
- *Closed loop obstruction:* this is a more ominous sign and is associated with distal small bowel obstruction ► the volvulus causes venous obstruction and small bowel necrosis – the small bowel loops will be thickened and oedematous (± pneumatosis) and any gas cannot be reabsorbed from the bowel lumen
- A gasless abdomen can be seen with prolonged vomiting, a closed loop obstruction with viable small bowel, or with a massive midgut necrosis

Upper GI study

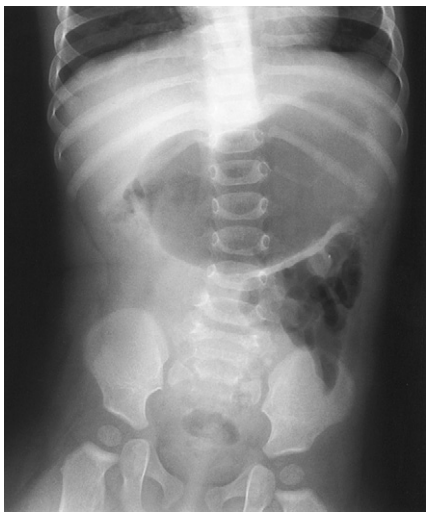
- **Normal:** on a supine AXR the normal duodenojejunal junction lies to the left of the left-sided pedicles at the height of the duodenal bulb ► on a lateral view, the junction of the 2nd and 3rd parts of the duodenum is retroperitoneal
- **Malrotation:** the duodenojejunal junction is displaced inferiorly and to the right on a supine AXR ► the junction of the 2nd and 3rd parts of the duodenum turns sharply anterior ► the distal jejunal loops lie to the right of the midline ► the caecal pole may lie high and more to the left side
- **'Corkscrew' pattern:** this describes the duodenum and jejunum spiralling around the mesenteric vessels and is pathognomonic for a midgut volvulus
  - If Ladd bands are causing the duodenal obstruction rather than a volvulus the duodenojejunal course has been described as 'Z-shaped' rather than spiral

US A dilated, fluid-filled stomach and proximal duodenum if obstruction is present ► the superior mesenteric vein (SMV) lies ventral or to left of the superior mesenteric artery (SMA) in about ⅔ of patients

- *'Whirlpool' sign:* the volvulus itself may be demonstrated (colour Doppler studies may show the SMV spiralling clockwise around the SMA)

PEARLS

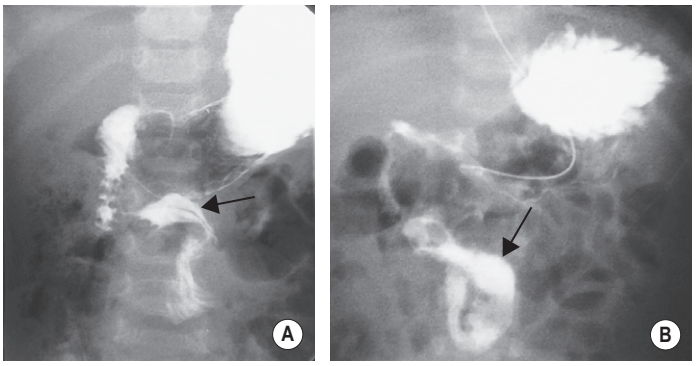
- Normal fetal gut development:
  - The gut begins as a straight, midline tube, which, as it elongates and develops, herniates into the base of the umbilical cord
  - Between the 6th and 10th weeks of fetal development, the midgut loop rotates 90° anticlockwise around the axis of the SMA ► at this stage the duodenojejunal (proximal) loop lies to the right, and the caecocolic (distal) loop lies to the left
  - During the 10th week, the intestines return to the abdominal cavity (the proximal loop of bowel enters first)
  - Both the proximal and distal loops undergo a further 180° of anticlockwise rotation as they return to the abdominal cavity (a total of 270° of rotation)
  - The duodenojejunal loop comes to lie posterior to and the caecocolic loop anterior to the SMA ► the duodenojejunal junction (fixed by the ligament of Treitz) should lie in the left upper quadrant of the abdomen, and the ileocaecal junction within the right lower quadrant
- If there is complete duodenal obstruction on an AXR or clinical peritonitis, a neonate requires surgery rather than an upper GI study
- The majority of patients have an isolated bowel malrotation, but there is an increased association with: duodenal stenosis and atresia ► omphalocele ► gastroschisis ► congenital diaphragmatic hernia ► heterotaxy syndromes ► Hirschsprung's disease ► megacystis–microcolon–intestinal hypoperistalsis (Berdon) syndrome ► congenital short bowel without atresia



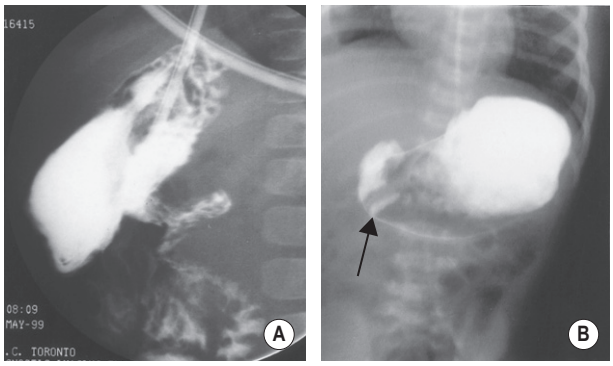
Malrotation and volvulus. AXR in a 12-month-old boy with bilious vomiting. The stomach is distended with a relative paucity of gas distally.†

Follow-through examination demonstrating an abnormal high and medial caecal position (arrow). Malrotation confirmed at surgery.†

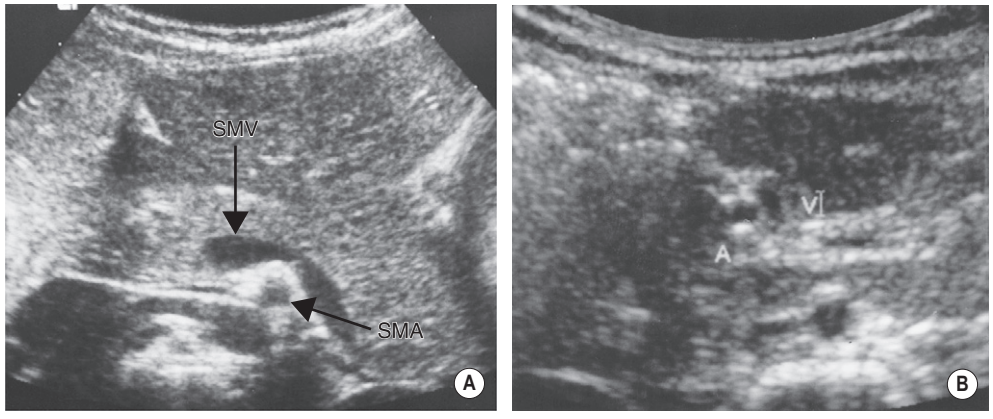
Ultrasound 'whirlpool' sign of midgut volvulus. Twisting mesenteric vessels with concentric rings of echogenic mesentery.†



Malrotation. Contrast meal (A) demonstrating abnormally low position of the duodenojejunal junction (arrow). Further case (B) demonstrating both inferior and medial displacement of the duodenojejunal junction (arrow). The normal junction should lie to the left of the midline (over or lateral to the left vertebral pedicle) at the level of the pylorus.†



Midgut volvulus presenting as a classic 'corkscrew' appearance of the duodenum and proximal jejunum on lateral view (A) and in a further case as complete duodenal obstruction (B). The linear filling defect (arrow) is likely to represent the superior mesenteric vessels and associated mesentery.†



Transverse ultrasound images of the normal SMA:SMV relationship (A) and a malrotated child (B) in whom the SMV lies to the left of the SMA.†

HIRSCHSPRUNG'S DISEASE

Definition

- A form of functional low bowel obstruction due to failure of caudal migration of neuroblasts within developing bowel – this results in an absence of parasympathetic intrinsic ganglion cells in both Auerbach's and Meissner's plexi ► the distal large bowel from the point of neuronal arrest to the anus is *continuously* aganglionic ('skip lesions' are rare)
  - *Short segment disease (75% of cases):* the aganglionic segment extends only to the rectosigmoid region ► short segment disease is sporadic (M > F)
  - *Long segment disease:* this involves a portion of the colon proximal to the sigmoid colon ► long segment disease has a strong familial incidence (M = F)
- Variants of Hirschsprung's disease include *total aganglionosis coli* and *total intestinal Hirschsprung's disease* ► *ultrashort segment disease* is rare and involves only the anus at the level of the internal sphincter

Clinical presentation

- Neonatal abdominal distension ► vomiting ► failure to pass meconium (with a delay > 48 h)
- It may unusually present later in childhood with chronic constipation and failure to thrive ► it can rarely present with an acute abdomen due to a colonic volvulus

AXR This typically shows a low bowel obstruction – commonly with colonic dilatation out of proportion to the small bowel ► absent rectal gas (a non-specific sign also seen with sepsis and NEC) ► retention of contrast medium above the sigmoid colon (>24 h – also non-specific)

- 5% of infants will have a pneumoperitoneum (occurring commonly within the ascending colon and which may be appendiceal)
- Intraluminal small bowel calcifications may be present with long segment disease
- A coexistent enterocolitis may lead to mucosal oedema, ulceration and spasm

Contrast enema (With a false-negative rate of 20–30%) ► a balloon catheter should not be used (the balloon can obscure the diagnostic features or even perforate the stiff aganglionic bowel)

- Lateral view:
  - *Short segment disease:* a narrow rectum with a cone-shaped transition zone to the more proximal, dilated and ganglionated bowel ► the radiological transition zone is commonly found distal to the pathological transition zone – in addition a transition zone may not be present in the neonate as it takes time for the proximal bowel to dilate
    - *Rectosigmoid ratio:* the rectum should always be the most distensible portion of the bowel with a rectosigmoid ratio > 1 ► this ratio is reversed in short segment disease
  - *Total aganglionosis coli:* the findings are unreliable ► there is shortening of a normal-calibre colon (with loss of the normal redundancy of the flexures) ► muscle spasm, a pseudo-transition zone, easy reflux of contrast medium into the terminal ileum, and a microcolon may also be seen

Pearls

- **Associations:** Down's syndrome (5%) ► ilial and colonic atresias ► cleft palate ► polydactyly ► craniofacial anomalies ► cardiac septal defects ► other neurocristopathies
- **Definitive diagnosis:** suction rectal biopsy

IMMATURE LEFT COLON (MECONIUM PLUG SYNDROME OR SMALL LEFT COLON)

Definition A relatively common cause of neonatal bowel obstruction with delayed passage of meconium due to inspissated meconium ► it is possibly due to immaturity of the myenteric plexus

- It is *not* associated with cystic fibrosis

Clinical presentation Symptoms and signs of bowel obstruction (patients tend to be less ill than those with a mechanical obstruction)

AXR Distended small and large bowel loops to the level of the inspissated meconium plugs ► a few fluid levels are seen

Contrast enema Typically there is a microcolon distal to the splenic flexure, at which point there is an abrupt transition to a mildly dilated proximal colon (in Hirschsprung's disease the transition zone is more gradual and is uncommon at the splenic flexure) ► the rectosigmoid ratio is normal

- Discrete plugs of meconium are seen as filling defects within the dilated colon
- There is a therapeutic (as well as a diagnostic effect) if water-soluble contrast medium is used – meconium is typically passed soon after

COLONIC ATRESIA

Definition A rare condition due to an in utero vascular accident (the right colon is commonly affected) ► the atresia may take on the form of a diaphragm or web, fibrous cord or mesenteric gap defect

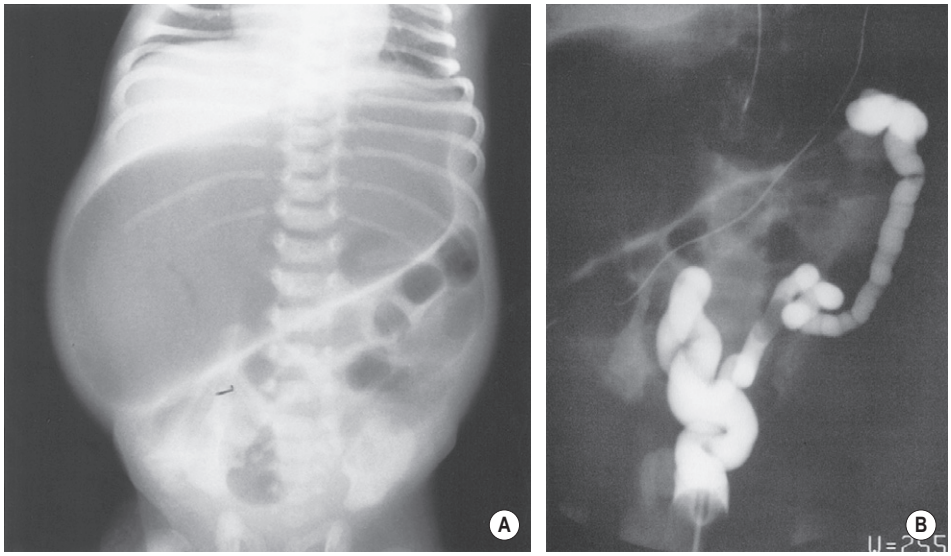
- It is associated with proximal atresias, gastroschisis, and Hirschsprung's disease

Clinical presentation Symptoms and signs of bowel obstruction

AXR Features of a low intestinal obstruction

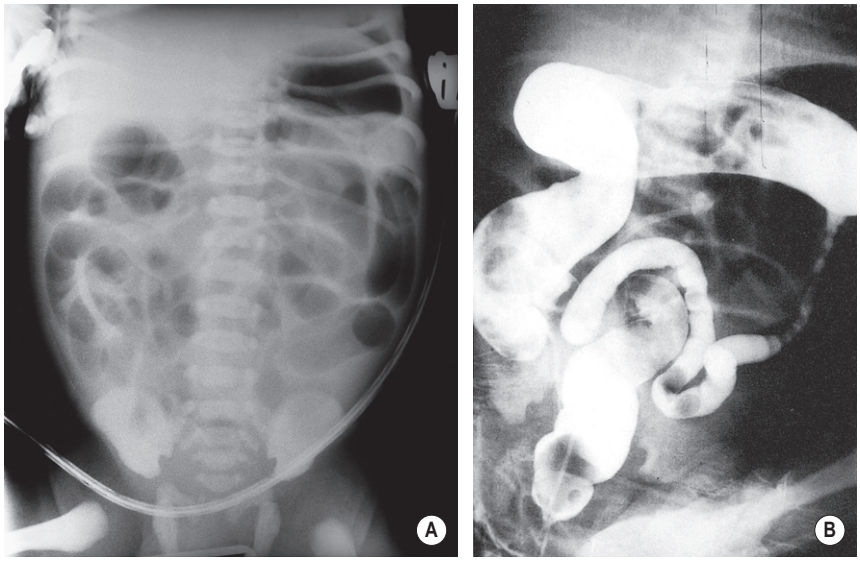
Contrast enema A distal microcolon, with obstruction to the retrograde flow of contrast medium at the point of the atresia

- *'Wind sock' configuration:* if a colonic diaphragm or web is present, the column of barium may cause the obstructing membrane to balloon into the proximal air-filled colon

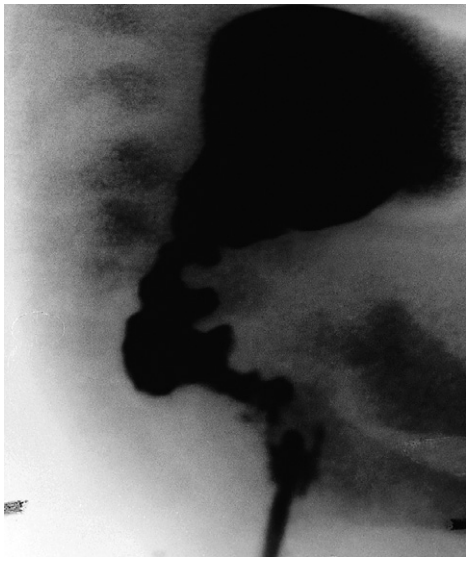


Colonic atresia. AXR (A) showing disproportionate dilatation of one bowel loop in a neonate with abdominal distension and failure to pass meconium. Contrast enema (B) showing a blind-ending colon with a convex distal border in the splenic flexure. The dilated air-filled proximal colonic segment can be seen. An isolated colonic atresia was confirmed at surgery.†

Functional immaturity (left colon syndrome). Contrast enema in a newborn term infant showing a relatively small left colon, transition zone at the splenic flexure and a large coiled meconium plug which was dislodged from the splenic flexure to the hepatic flexure during colonic filling.†



Small left colon syndrome. (A) Supine AXR shows a low obstruction with multiple dilated loops of bowel. (B) Contrast medium enema shows a microcolon distal to the splenic flexure. The transition point is abrupt.*



Rectosigmoid Hirschsprung's disease. Lateral view, contrast medium enema. The cone-shaped transition zone, abnormal rectosigmoid ratio and tertiary rectal contractions are demonstrated.*

ABDOMINAL MANIFESTATIONS OF CYSTIC FIBROSIS

The GI complications of cystic fibrosis result from abnormally viscous secretions within the hollow viscera and the ducts of the solid organs.

Meconium ileus

Definition This is a form of distal intestinal obstruction caused by inspissated pellets of meconium within the terminal ileum ► > 90% of patients have cystic fibrosis (meconium ileus is the presenting feature of cystic fibrosis in 10–15%)

Clinical presentation Vomiting ► abdominal distension ► failure to pass meconium

Complications Intrauterine volvulus (due to a heavy, meconium-laden loop of bowel) ► a volvulus can lead to stenoses, atresias and perforation

- Perforation leads to a chemical meconium peritonitis with subsequent fibrosis and calcification
- Other causes of meconium peritonitis include a small bowel atresia or an intrauterine intussusception

AXR

- **Uncomplicated ileus:** small bowel dilatation ► fluid levels are scant
  - *A 'soap bubble' appearance:* this is due to an admixture of meconium with gas ► (classically seen within the right iliac fossa)
- **Complicated ileus:** intra-abdominal (bowel wall) or scrotal calcification ► prominent air-fluid levels
  - *A meconium pseudocyst:* this occurs due to a vascular compromise in association with an intrauterine volvulus ► the ischaemic bowel loops become adherent and necrotic, and a fibrous wall develops around them ► the wall may then calcify and the cyst can have a secondary mass effect

US

- **Uncomplicated ileus:** dilated bowel loops are filled with echogenic material (cf. echo-poor material with an ileal atresia)
- **Complicated ileus:** 'snow storm' ascites with a meconium peritonitis

Contrast enema There is a virtually empty microcolon ► reflux of contrast medium is seen into a small terminal ileum (with numerous pellets of meconium outlined) ► there are proximally dilated mid-ileal loops

Pearls

- A Gastrografin enema may be therapeutic in an uncomplicated case – Gastrografin is hypertonic and will draw water into the bowel lumen by osmosis, softening the meconium and allowing it to pass ► there is a risk of perforation (5%) or a fluid–electrolyte imbalance (therefore only use half-strength Gastrografin in a well-hydrated infant)
- Bowel obstruction in neonates with cystic fibrosis can be due to either meconium ileus or meconium plug syndrome

Distal intestinal obstruction syndrome

Definition Impaction of mucofeculent material within the terminal ileum and right colon (which is seen in 10–15% of older children with cystic fibrosis) ► it is potentially fatal

Clinical presentation Colicky abdominal pain and distension ► nausea and vomiting ► constipation ► a right iliac fossa mass

AXR Faecal loading of the colon with a 'bubbly' appearance ► right-sided abdominal mass ► dilated small bowel

Treatment Oral Gastrografin (± a Gastrografin enema) to soften and mobilize the stool

Fibrosing colonopathy

Definition A colonic stricture due to the irreversible and sometimes progressive narrowing of the bowel lumen with associated submucosal fibrosis and fatty infiltration ► it often involves the right side of the colon

- High-strength pancreatic enzyme supplements have been implicated in its aetiology

Clinical presentation Distal intestinal obstruction

AXR/MRI/US Thickened bowel wall

Contrast enema Shortening of the colon with narrowing of the colonic lumen ► loss of the colonic haustration ► nodular thickening of the colonic wall

Treatment Surgical resection

Pancreatic insufficiency

Definition This occurs in 80–85% of children with cystic fibrosis and manifests as malabsorption (chiefly of fat and proteins) ► 30–50% of patients have glucose intolerance (with 1–2% requiring insulin therapy)

AXR Punctate calcification within the pancreas

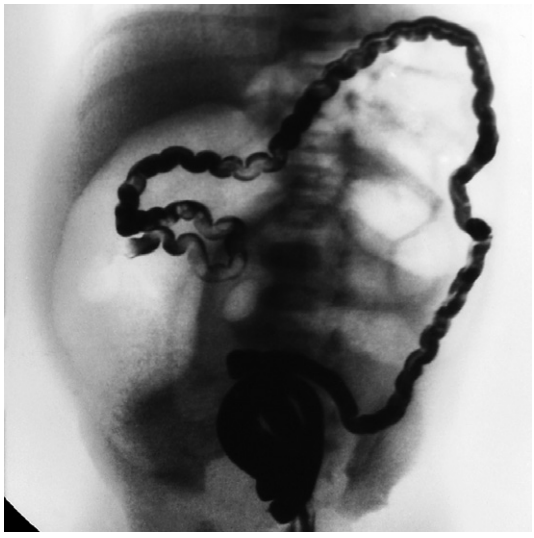
US A small echogenic pancreas

CT/MRI Fatty replacement of the pancreas

Miscellaneous

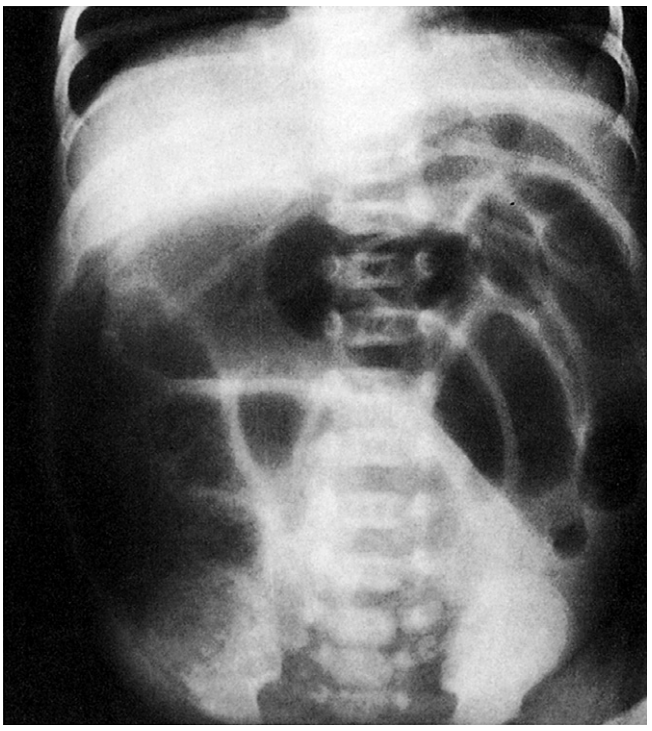
- **Liver cirrhosis:** this results from impaired biliary drainage with associated portal hypertension (there can be splenomegaly and gastric varices)
- **Chronic cholecystitis:** there is a small, thick-walled gallbladder (gallstones are seen in 10% of patients)
- **Intra-abdominal malignancy:** cystic fibrosis is associated with cancers of the oesophagus, stomach, small bowel, colon, liver, biliary tract, pancreas and rectum
- **Other bowel manifestations:** peptic ulcer disease ► GOR ► oesophagitis ► oesophageal stricture ► thickened nodular mucosal folds within the duodenum and small bowel
  - Intussuception: this occurs in 1% of patients (usually between the ages of 9–12 years) ► it is usually ileocolic)

Fatty replacement of the pancreas in a patient with cystic fibrosis.••

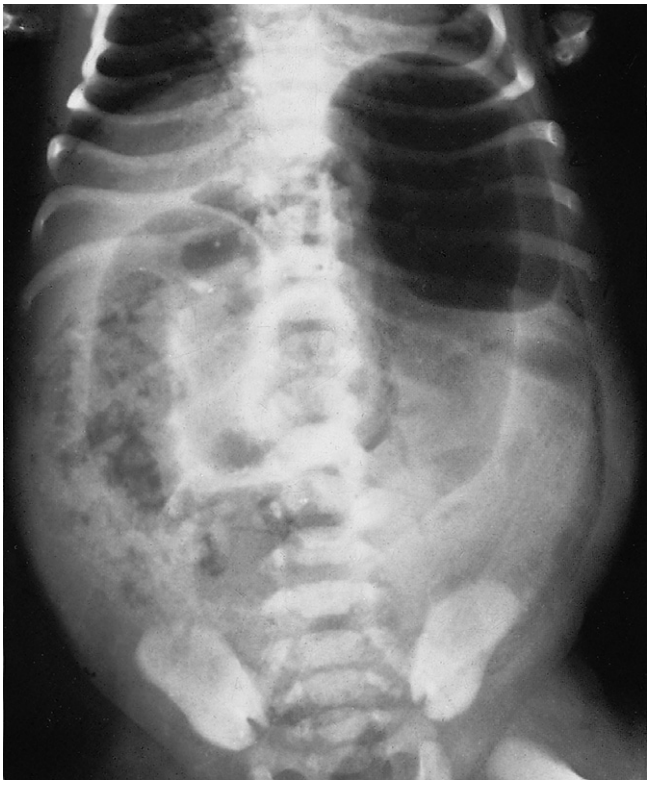


Meconium ileus. Contrast enema demonstrates the empty microcolon. Contrast refluxes into the narrow terminal ileum, where pellets of meconium are outlined.*

Meconium ileus. Contrast enema demonstrating a microcolon with reflux into dilated distal ileum. Multiple filling defects of inspissated meconium are seen within the distal ileum (superimposed over the transverse colon) and the colon.†



Meconium ileus. Supine AXR shows a low obstruction with multiple, dilated loops of bowel and a 'soap bubble' appearance in the right lower quadrant.*



Meconium ileus. AXR showing loops of dilated bowel with a 'bubbly' appearance of meconium mixed with air in the right side of the abdomen. Free air is seen, indicating a perforation.†

NECROTIZING ENTEROCOLITIS

DEFINITION

- This is an often severe enterocolitis that affects primarily premature infants (with an increasing incidence due to the increased survival rates of very low birth weight infants of a younger gestational age)
  - NEC can also be seen in term infants (particularly those with polycythaemia, cyanotic congenital heart disease and gastroschisis)
- Initially superficial, the inflammatory process can extend to become transmural ► diffuse or discrete involvement of the bowel can occur ► the most commonly affected sites are the terminal ileum and colon (up to 50% of cases involve both the small and large bowel)
- The aetiology remains unknown, but immaturity of the gut mucosa and immune response (coupled with ischaemia and hypoxia) are felt to contribute ► there is also a possible infectious cause
  - *Additional risk factors:* sepsis ► early enteral feeding ► umbilical arterial and venous cannulation ► maternal cocaine abuse
    - Breastfeeding is associated with a decreased risk of developing NEC

CLINICAL PRESENTATION

- The initial clinical symptoms and signs are non-specific: lethargy ► hypoglycaemia ► temperature instability ► bradycardia ► feeding intolerance ► increased gastric aspirates ► gastric distension
  - Disease progression leads to vomiting and diarrhoea (often with the passage of blood or mucus in the stool), and eventually to shock
  - Severely affected infants may have visibly erythematous anterior abdominal walls, with palpable distended loops of bowel
- Perforation will occur in ⅓ of children and occurs most commonly in the ileocaecal region (affecting 60% of cases)

RADIOLOGICAL FEATURES

AXR An early sign is diffuse gaseous distension of both the small and large bowel (or isolated gastric distension) ► serial XRs (taken every 6–12 h) will demonstrate fixed bowel loop dilatation and thickening (oedema) with loss of distinction of the bowel walls

- If the diameter of a bowel loop is greater than the width of the L1 vertebral body then it is likely to be dilated
- **Intramural gas (pneumatosis intestinalis):** this is a more specific sign, and an increasingly extensive pneumatosis correlates with an increased NEC severity
  - *Submucosal gas:* 'bubbly' lucencies in the bowel wall
  - *Subserosal gas:* linear bowel wall lucencies
- **Portal venous gas:** this is seen in approximately 10% of cases and is associated with severe NEC (its presence does not necessarily imply a fatal outcome) ► the disappearance of intramural or portal venous gas may herald imminent perforation rather than recovery
- **Indicators of imminent perforation:** free intraperitoneal fluid ► the 'persistent loop' sign: a solitary, dilated loop of bowel present over 24–36 h
- **Perforation:** < ⅔ of patients will have free air visible on a plain XR (almost all patients who perforate will do so within 30 h of diagnosis)
  - *'Football' sign:* a large elliptical central abdominal lucency in the supine position (due to rising intra-abdominal air)
  - *'Telltale triangle' sign:* the collection of small amounts of free intraperitoneal air between loops of bowel seen with a supine, cross-table lateral or decubitus view

Contrast enema Its use in the acute situation (if the other signs are ambiguous) is controversial due to the risk of sepsis and perforation

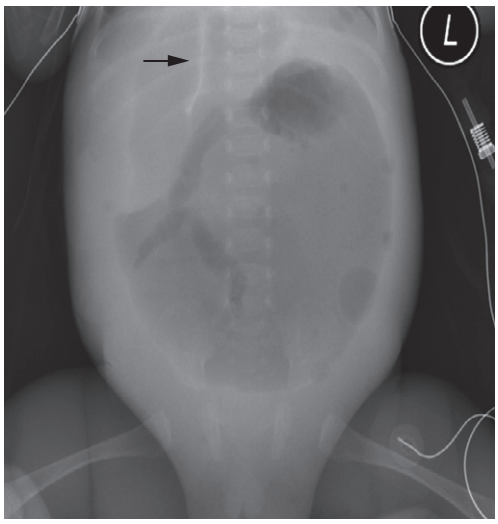
US This is more sensitive than an AXR in the detection of ascites and portal venous gas

- **Portal venous gas:** echogenic particles flowing within the portal vein or focal areas of intrahepatic increased echogenicity
- **The 'circle sign':** this is indicative of bubbles of gas circumferentially within the bowel wall and is seen as a continuous, echogenic ring in cross-section
- **Perforation:** the presence of free intraperitoneal fluid may be an indicator of perforation (it is seen in only 20% of patients)

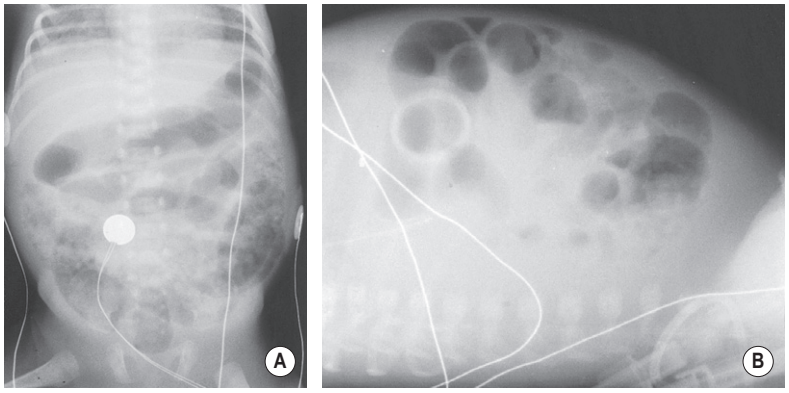
PEARLS

- The overall mortality rate from NEC is approximately 30%
- Perforation is not an absolute indication for surgical intervention – peritoneal drains are used in the initial resuscitation (delaying the need for surgery and allowing time for systemic recovery) ► in some instances a peritoneal drain may provide definitive treatment
- **Complications:** a late complication is stricturing, which can be single or multiple, and occurs in up to ⅓ of patients ► the majority of strictures are short, are found in the colon, and are diagnosed up to 3 months following the acute illness
  - *Other late complications:* an acquired intestinal atresia (rare) ► abscess formation ► enteric fistulas ► enterocyst formation ► obstruction secondary to adhesions ► malabsorption ► short bowel syndrome following surgical resection

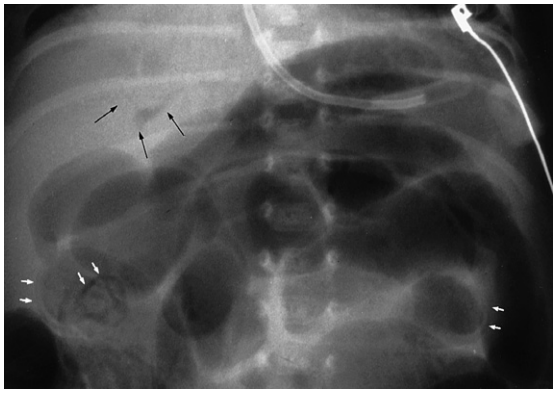
Necrotizing enterocolitis. Supine AXR demonstrates multiple dilated loops of bowel, extensive submucosal and subserosal pneumatosis, and portal vein gas.*



The 'football' sign: free intraperitoneal air has collected superiorly in this supine AXR, creating a central lucency. The free air is also outlining the falciform ligament (arrow).

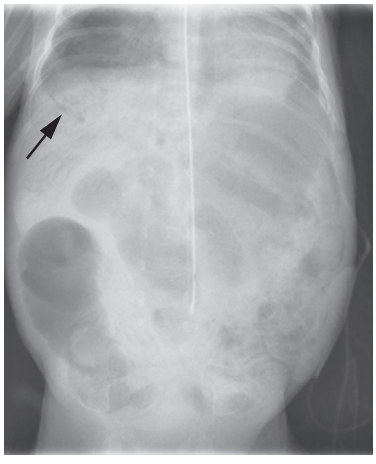


Necrotizing enterocolitis. Supine AXR (A) demonstrating extensive 'bubbly' pneumatosis in a 16-day-old premature infant born at 28 weeks' gestation. No definite free air seen. However, lateral shoot-through XR (B) shows a small triangle of free air beneath the anterior abdominal wall (arrow). A localized ileal perforation was found at laparotomy.†



Necrotizing enterocolitis with portal venous air. Branching lucencies throughout the liver represent gas within the portal venous system (*black arrows*). Intramural air is also present (*white arrows*).©20

NEC. Supine AXR demonstrating gas within the hepatic portal veins (arrow), together with subserosal bowel wall gas (best seen in the left lower quadrant).



Free intraperitoneal air in perforated necrotizing enterocolitis demonstrated by lucency over the entire abdomen ('football' sign), subdiaphragmatic air and outlining of both sides of the bowel wall (Rigler's sign).†

ANORECTAL MALFORMATIONS

DEFINITION

- Anorectal malformations include anorectal atresia and an imperforate anus (± an anomalous connection between the atretic anorectum and the genitourinary tract) ► it results from the failure of descent and separation of the hindgut and the GU tract during the 2nd second trimester (affecting 1 in 1500–5000 live births)
  - *A high lesion:* the rectum ends above the puborectalis sling
  - *A low lesion:* the rectum ends below the puborectalis sling
- It is associated with Down's syndrome (2–8%) as well as:
  - *The VACTERL sequence* (45% of patients)
  - *The OEIS complex* (5% of patients): **O**mphalocele + bladder **E**xstrophy + an **I**mperforate anus + **S**acral anomalies
  - *Currarino's triad:* an anorectal malformation (commonly anorectal stenosis) + bony sacral anomalies (classically a 'scimitar sacrum' with unilateral hypoplasia of the lateral aspect of the vertebral bodies) + a presacral mass lesion (e.g. an enteric cyst, teratoma, anterior meningocele or dermoid)

CLINICAL PRESENTATION

- **Low lesions:** usually there is a visible perineal opening ► the orifice may be located more anteriorly than normal (an ectopic anus) and it may be stenotic or covered with a membrane ► there is no communication with the GU tract ► low lesions also include an isolated rectal atresia or stenosis
  - Female patients with low lesions will have separate urethral and vaginal orifices with an intact hymen
- **High lesions:** there is no visible perineal fistula ► rarely the rectum ends blindly
  - Male patients will usually have a fistulous tract between the atretic anorectum and the posterior urethra (less commonly a fistula to the bladder or anterior urethra)
  - Female patients usually have fistulas from the atretic anorectum to the vagina or vestibule

RADIOLOGICAL FEATURES

Inverted lateral XR A radio-opaque marker is placed over the anal dimple and the distance between the pouch of rectal gas and the marker is measured

- *False positives:* if a patient is imaged during the 1st 24 h of life (as any gas may not have yet reached the rectum) ► if the infant has not been held prone for long enough ► if meconium has impacted within the distal rectum
- If an infant is crying or straining, the rectal pouch can descend through the levator sling and a high lesion may be misinterpreted as a low one

Supine AXR This can detect any associated bony anomalies of the spine

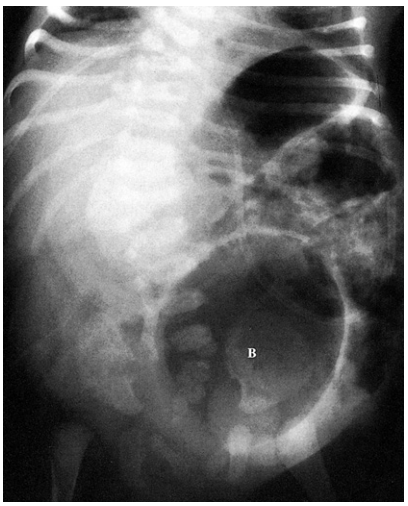
- *Intravesical air:* this implies a high lesion (with a rectovesical fistula or a rectourethral fistula in a boy)
- *Calcified intraluminal meconium:* this implies a high lesion in a boy (meconium calcifies when it comes into contact with urine)

Transperineal US This measures the distance of the rectal pouch from the perineum ► there are problems with interpretation as for an inverted lateral XR

Augmented pressure colostogram This is performed in infants with high lesions following the initial formation of a colostomy ► a Foley catheter is inserted into the distal segment of colon and a balloon is inflated to 5ml ► water-soluble contrast is then hand injected to distend the distal colon and define the fistulous tract

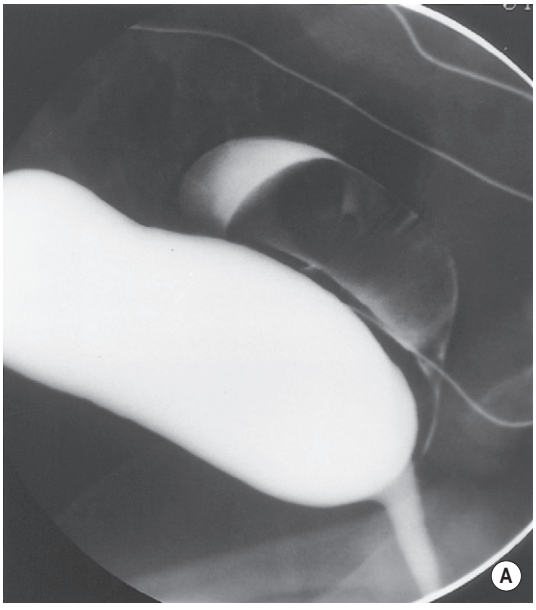
PEARLS

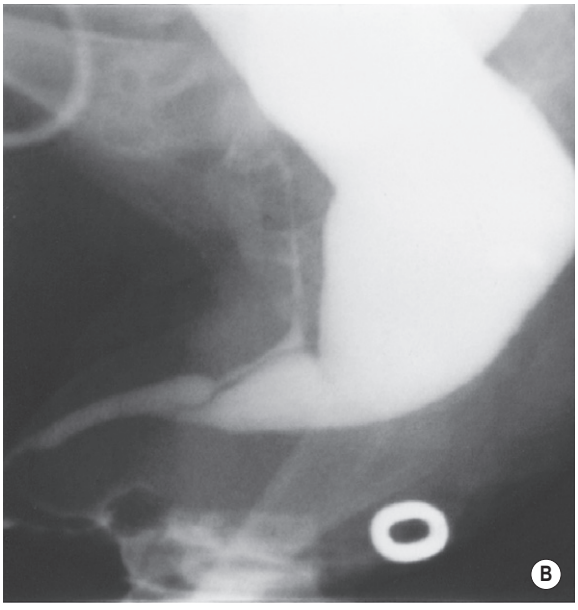
- Renal US is mandatory in all infants with anorectal atresia ► a spinal US will exclude any associated spinal cord lesions (e.g. cord tethering) as these are not uncommon associations
  - Alternatively a pre- and postoperative MRI can study the pelvic floor and reveal any associated renal or spinal anomalies
- **Low lesions:** these are treated with an anoplasty or dilatation soon after birth
- **High lesions:** these are treated with a colostomy and then definitive repair



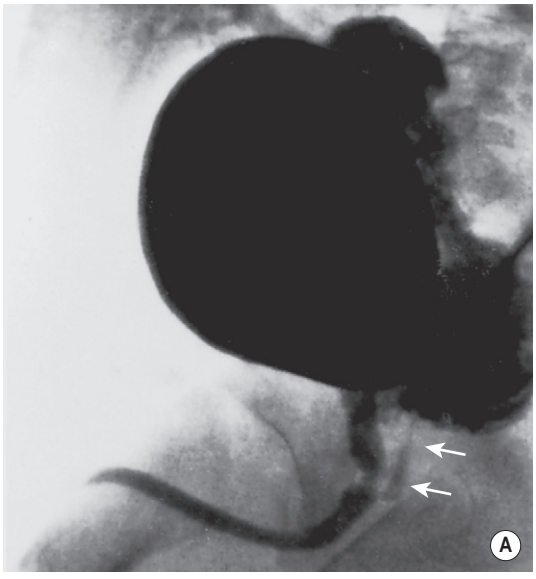
Colovesical fistula. Supine AXR shows intravesical air and vertebral segmentation anomalies in a male infant with a high anorectal malformation. B = bladder.*

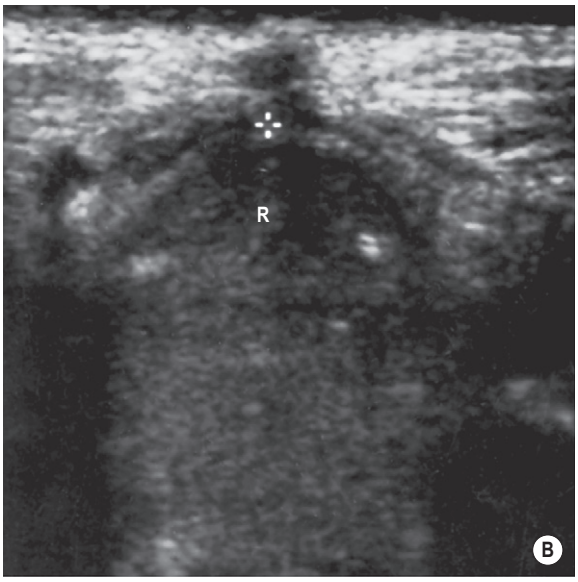
Recto-urethral fistula. Augmented pressure colostogram in a male infant with a high anorectal malformation.*





Anorectal atresia with urethral fistula. Micturating cystogram (A) demonstrating fistula from the distal rectal pouch to the posterior urethra and distal loopogram. (B) showing a fistula to the anterior urethra.†





Imperforate anus. (A) Communication between the posterior urethra and the rectum (arrows). (B) Transperineal US scan illustrates the distance from the pouch (R) to the anal dimple (marker) as well as calcific densities within the meconium within the rectal pouch.§

NEUROBLASTOMA (NB)

DEFINITION

- A malignant tumour arising from primordial neural crest cells (these normally develop into the adrenal medulla and sympathetic ganglia) ► tumours can arise from the adrenal glands or usually anywhere along the sympathetic chain within the neck, thorax and abdomen
  - *Thorax*: tumours arise within the posterior mediastinum
  - *Abdomen:* tumours arise within the retroperitoneum in 60% of cases, and of these, 40% occur within the adrenal glands
  - It is the 2nd commonest paediatric abdominal cancer (after Wilms' tumour)
- **Classification of neural crest tumours:**
  - *Neuroblastoma:* malignant
  - *Ganglioneuroblastoma:* intermediate features
  - *Ganglioneuroma:* well differentiated and benign

CLINICAL PRESENTATION

- The median age at diagnosis is 2 years (90% are diagnosed at < 5 years of age)
- It presents with an incidental mass or abdominal pain ► other presenting features:
  - *Symptoms due to metastatic disease:* bone and joint pain ► proptosis (orbital metastases) ► anaemia ► weight loss ► fever
  - *Excess hormone production:* hypertension (catecholamines are produced in 95% of patients) ► intractable watery diarrhoea (VIP production)
  - *Horner's syndrome:* this follows involvement of the stellate ganglion

RADIOLOGICAL FEATURES

CXR A calcified mass ► paravertebral widening within the lower chest (due to any retrocrural spread)

AXR A non-specific soft tissue mass with calcification seen in up to 50% of cases

Long bones Metastases can appear as ill-defined areas of bone destruction ► a solitary lesion may appear as a lytic, moth-eaten or permeating destructive area interspersed with sclerotic trabeculae ► there may be a laminar periosteal reaction

- Common sites are the skull and long bone metaphyses ► they are not seen within the hands, feet, or clavicles

US A heterogeneous and hypervascular mass of varying echogenicity ► hypoechoic areas are secondary to haemorrhage and necrosis

US/cross-sectional imaging This determines the local disease extent

- An aggressive large and heterogeneous mass which invades the adjacent structures and readily crosses the midline ► it tends to surround and engulf (rather than displace) large vessels ► there will be external compression on the kidney ► the tumour may invade the spinal canal, kidney or liver
- *Skull metastases:* these appear as an infiltrating mass causing permeative bone destruction and spiculated bone changes – these may extend into the scalp soft tissues or push through the skull inner table

CT Calcification is seen in up to 85% of cases (diffuse, mottled, finely stippled, or coalescent) ► erosion of the pedicles is suggestive of intraspinal extension ► low attenuation areas represent areas of haemorrhage and necrosis ► mild heterogeneous enhancement reflects areas of vascularity alternating with areas of necrosis, haemorrhage and cystic change

MRI T1WI: a low SI mass ► T2WI: a high SI mass ► T1WI (FS) + Gad: heterogeneous enhancement

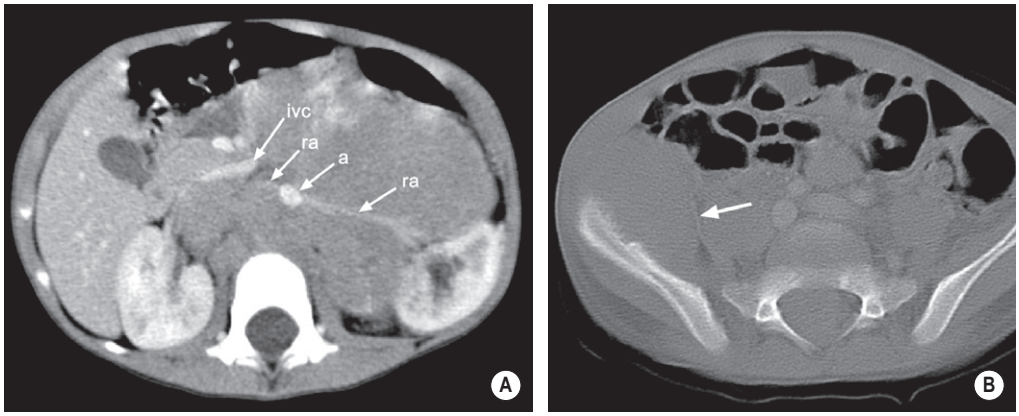
- *Extradural extension (dumb-bell syndrome):* this is common with thoracic NB, but rare with abdominal tumours
- *Bone metastases:* T1WI: low SI ► T2WI: high SI

Scintigraphy This determines the distal disease extent

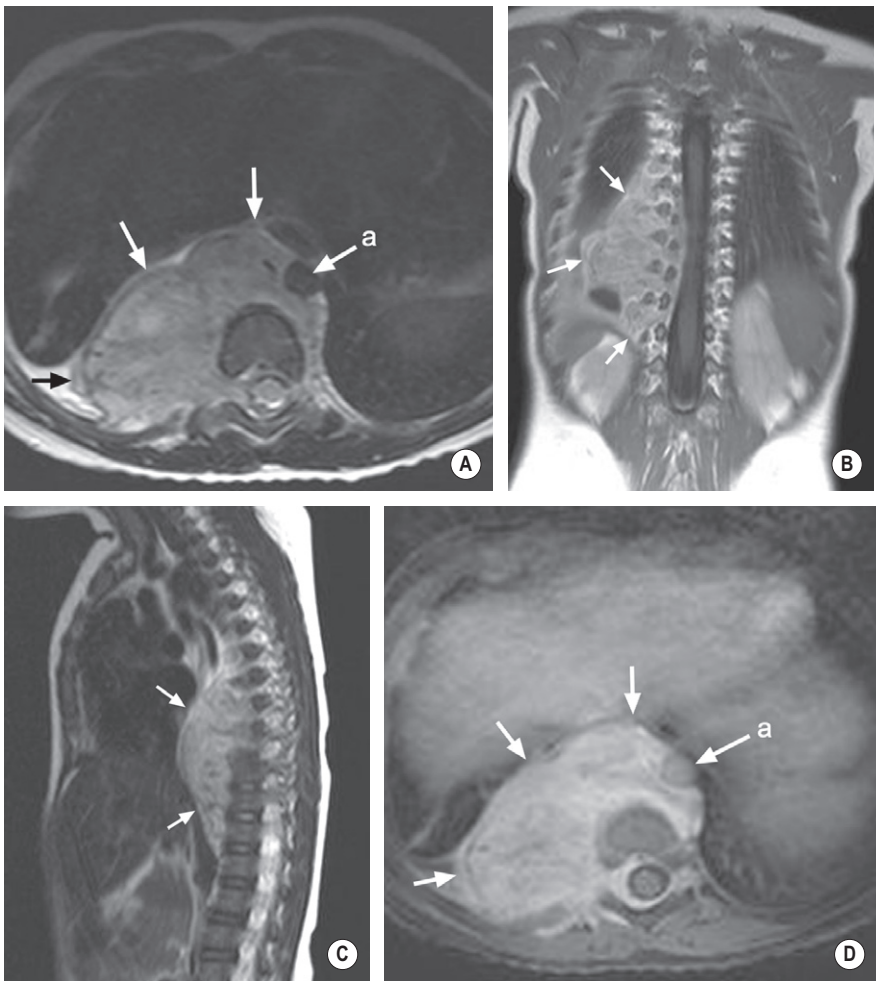
- **MIBG** (^{131}I- or ^{123}I-labelled): an analogue of the catecholamine precursors taken up by catecholamine-producing cells ► it is usually specific for NB (primary tumour and metastases) although 30% of primary lesions do not take up MIBG
- **^{99m}Tc-MDP bone scintigraphy:** this should be performed in all patients at diagnosis and follow-up as ⅔ of patients have metastases at presentation
- **FDG PET:** this has a limited role ► it demonstrates a similar imaging pattern as with MIBG (diffuse abnormal skeletal uptake with extensive bone marrow involvement) ► it cannot visualize lesions in the cranium because of the high physiological activity within the brain

PEARLS

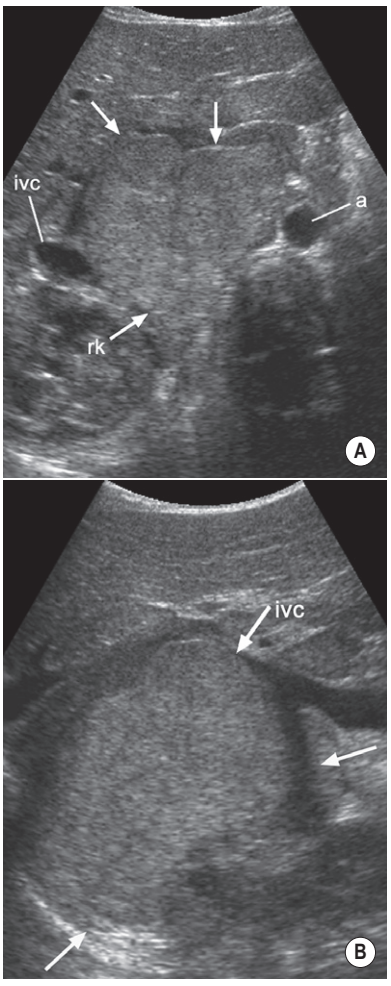
- Neuroblastoma tends to regress in size with treatment but the regression is frequently incomplete (leaving a small residual soft tissue mass which is often calcified) ► MIBG is required to determine whether this residual tissue is fibrosis or viable tumour
- **Staging (see Section 9):**
  - ⅔ of patients have metastatic bone disease at diagnosis
    - *Involved sites:* the skeleton ► bone marrow ► lymph nodes ► liver ► rarely the lung and brain
- **Stage IV-S:** this has a better prognosis with a tendency to spontaneously regress
  - The hepatomegaly may be massive despite a small primary lesion (and may cause severe respiratory complications in very young patients) ► there may be bone marrow lesions and palpable subcutaneous nodules



Abdominal neuroblastoma. CECT. (A) A prevertebral tumour extends across the midline in the retroperitoneum, displaces and encases the aorta (a) anteriorly and encases the renal arteries (ra). The inferior vena cava (ivc), partially encased, is displaced anterolaterally and compressed by the tumour and nodes. The mass extends to both the renal hili. (B) Infiltrative lesion to the right iliac bone with a large soft tissue component (arrow) that projects inwards as a space-occupying mass.*



Thoracic neuroblastoma with intraspinal extension. MRI. (A,B) Axial and coronal T1WI + Gad. (C) Sagittal T1WI + Gad. (D) Axial T2WI. Large posterior mediastinal mass (arrows) with anterior and lateral displacement of the aorta (a), which is encased. There is massive tumour extension into the canal through the right neural foramina to displace the spinal cord to the left. Extension into the right paraspinal soft tissues is evident.*



Abdominal neuroblastoma. (A) Transverse US through the right abdomen shows a solid paraspinal mass (arrows) anterior to the right kidney (rk). The aorta (a) and inferior vena cava (ivc) are displaced by the mass. (B) Longitudinal image through the right flank shows the mass (lower arrows) and the stretched inferior vena cava (ivc).*